Radiology of the Liver in Children

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Introduction

Imaging can play an integral role in the evaluation, diagnosis, and even treatment of children with new or chronic hepatobiliary disorders involving a native or transplanted liver. The primary imaging modalities that are routinely called upon to evaluate the pediatric liver include ultrasound (US), computed tomography (CT), magnetic resonance imaging (MRI), and nuclear scintigraphy. The detailed anatomical capabilities offered by US, CT, and MRI have essentially eliminated the need for conventional x-ray images.

The appropriate and efficient utilization of the diverse diagnostic imaging modalities available is of great interest in order to maximize the diagnostic yield while limiting the potential side effects or risk to the child [1]. Each imaging study has its limitations and frequently multiple imaging modalities are necessary. The information garnered from various imaging studies can be complementary not only to one another but also further serve to complement the clinical exam along with various laboratory tests.

Ultrasound (*US*) is the imaging modality most frequently called upon for the initial screening evaluation of children with liver disorders. There are several reasons why sonography is quite appealing in the evaluation of the pediatric population. First, US permits real-time and diverse multi-planar imaging capabilities. Furthermore, Doppler US techniques provide the ability to characterize vascular flow in real time which can be critical in circumstances such as liver transplants. Second, US does not expose the child to ionizing radiation. Children from both their younger age and longer lifetime expectancy are more vulnerable to radiation-induced cancers than adults [2]. US offers the opportunity to follow longitudinally various conditions that may affect the child's liver with sequential follow-up examinations. Finally, it is a readily available modality that is also portable. This permits its utilization when the patient cannot ideally be transported to the imaging department.

Despite US having very appealing characteristics, it does have limitations. Although the spatial resolutions can be quite exquisite, the acoustic windows needed for imaging can be limited by overlying bandaging, bowel gas, and patient body habitus. Probably the greatest limitation of US is that it is very much operator dependent. It is critical that the sonographer, radiologist, and pediatric specialist requesting the study effectively communicate the expectation and

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limitations of the US imaging studies to maximize the utility of imaging interpretation.

Computed tomography (*CT*) has a valuable role in the imaging evaluation of children with liver disease. The modern multi-detector CT scanner offers the ability to produce multi-planar cross-sectional imaging studies with exquisite anatomical spatial resolution. The technology is readily available; studies can be rapidly performed without the need for sedation or anesthesia and are reliably reproducible. These reasons may account for the increased utilization of CT in children [3]. Given that CT exposes a child to ionizing radiation, its use should be limited to appropriate clinical indications, as with any medical test or study. CT imaging protocols that set parameters to optimize image production quality (adequate resolution) weighed against radiation exposure dose to the child should be taken into account by the radiology service providers. This is of special importance when children are imaged by providers that also routinely image adult patients and thus should adjust to pediatric appropriate imaging settings [3, 4].

Magnetic resonance imaging (*MRI*) has the capability of providing comprehensive evaluation of the liver parenchyma, biliary system, and vasculature. The imaging protocols can be tailored to optimize the evaluation of the liver to address specific clinical goals and imaging needs. Therefore, it is critical that the referring specialist and radiologist communicate prior to image acquisition in order to appropriately protocol the imaging sequences to be performed and maximize the information that can be acquired. Hepatic tumors are especially well suited to be evaluated by MRI in children with the goal of tumor characterization and assess the appropriateness of resection and tumor staging. The imaging characterization of liver tumor compared to normal hepatic parenchyma both before and following contrast enhancement is able to provide greater tissue differentiation than available with CT or US [5]. The multi-planar capabilities permit anatomical localization similar to that of CT with the added advantage of not exposing the child to ionizing radiation. The relative lack of operator dependence is an additional advantage compared to that of US. The primary limitation of MRI is often the time required to complete a comprehensive contrast study, which can be upwards of 1 h. As a result young children frequently require sedation or even anesthesia.

Nuclear scintigraphy is useful for the physiologic imaging of the pediatric hepatobiliary system. Iminodiacetic acid (IDA) binds readily to Technetium-99m and is excreted through the biliary system and thus the physiologic excretion pathway can be imaged. Normal IDA scan studies will demonstrate homogenous uptake throughout the liver typically within 5 min with subsequent clearance and excretion into the biliary ductal system. The gallbladder will retain tracer which can be identified along with excretion into the duodenum. In circumstances of biliary obstruction, the normal flow of radiopharmaceutical will not occur along with delayed hepatic uptake and clearance. In cases of suspected biliary leak, the radiotracer will accumulate in an abnormal extrabiliary location and single positron emission computed tomography (SPECT) can often clarify the location of the radiotracer material in difficult cases. Conditions with underlying hepatocyte dysfunction or absence of normal hepatocytes will have delayed or absent radiotracer uptake to the affected region. Technetium-99m sulfur colloid is a radiotracer that is not excreted into the biliary system. Its uptake is related to the normal function of hepatocytes during phagocytosis. As a result the distribution reflects the function of the reticuloendothelial cells of the liver and hepatic perfusion distribution.

Pediatric Liver Masses

Benign Tumors

One-third of primary tumors affecting children are benign and of mesenchymal or epithelial origin [6]. In a large series from the Armed Forces Institute of Pathology of liver tumors in patients under 21 years of age, infantile hemangioendothelioma was the most common benign primary hepatic tumor followed by focal nodular

hyperplasia (FNH), mesenchymal hamartoma, regenerating nodules, then hepatocellular adenoma [7].

Infantile hemangioendothelioma (*IH*) (also known as infantile hemangioma) is a benign (biologically) vascular neoplasm of infancy. They are most frequently diagnosed in the first 6 months of life, and one third will be diagnosed within the first month and manifests as an asymptomatic abdominal mass. However, serious complications can arise as a result of the hypervascularity and shunting effects such as congestive heart failure, Kasabach-Merritt syndrome, hypothyroidism, and rarely tumor rupture into the peritoneum.

IH can be solitary or multifocal and US findings can be suggestive but are not typically diagnostic. The main role of US is in the initial detection and for follow-up. When multifocal in the liver, there may also be involvement of other organs to include the chest and brain. Multifocal lesions tend to be small and uniform in appearance by US. Large focal or solitary lesions with high flow by Doppler US due to the hypervascularity can demonstrate enlargement of the hepatic arteries and veins with associated downstream tapering of the aorta below the celiac trunk Fig. 5.1.

MRI can offer confident diagnosis with characteristic findings before and after intravenous contrast. Pre-contrast tumors are generally well delineated from adjacent normal liver parenchyma. Smaller lesions tend to have a homogenous appearance and larger masses can have heterogeneous signal intensity. The lack of ionizing radiation permits the opportunity to perform multiple imaging phases following IV contrast to identify the enhancement characteristics that are typical of IH. The enhancement pattern of a large tumor typically demonstrates intense peripheral enhancement during the arterial phase with progressive central enhancement of the tumor during the portal venous phase. Smaller multifocal tumors can enhance intensely uniformly Fig. 5.2.

Multiphase dynamic contrast-enhanced CT can also be diagnostic of IH, but the need for ionizing radiation should limit this modality's use in these circumstances. Pre-contrast images will show a hypodense well-circumscribed mass compared to normal liver. In large lesions, speckles of calcifications are not uncommon. As with MRI, following the administration of IV contrast, there will be peripheral nodular enhancement during the arterial phase with progressive filling on delayed imaging phases Fig. 5.3.

Catheter angiography is no longer routinely used for diagnosis of IH but rather for percutaneous endovascular interventions. Embolization of the feeder arteries can be performed to slow down or occlude the hyperdynamic flow pattern supplying the tumor in children that develop clinical complications. Angiographic findings will show an enlarged hepatic arterial supply and dilated early draining veins.

Mesenchymal hamartoma is the second most common benign liver tumor occurring in children. They have imaging features that differ from IH. Diagnosis is dependent on pathologic findings, which can range from a predominantly complex heterogeneous mass with internal septations to a solid-appearing mass. Doppler US will show little vascularity except in the septations. Imaging by CT and MRI will demonstrate enhancement of only the internal septa and solid portions. The cystic portions will have similar characteristics as other fluid-filled structures in the abdomen such as the gallbladder or bladder. If predominantly solid, it will have a hypodense (by CT) or hypointense (by MRI) enhancement pattern following contrast Fig. 5.4.

Focal nodular hyperplasia (*FNH*) is a benign epithelial liver tumor composed of hepatocytes, Kupffer cells, vascular structures, and biliary ducts. Although most commonly identified in adult women, it can occur in young children and adolescents. The imaging appearance reflects the pathologic composition, very similar to the normal liver. By US an FNH can appear as a homogenous, well-circumscribed mass that is isoechoic, hypoechoic, or even hyperechoic to that of adjacent normal liver parenchyma. The "central scar" will be hyperechoic with increased vascularity compared to the rest of the mass. On unenhanced CT an FNH is well circumscribed and can be isodense or slightly hypodense relative to normal liver. Following IV contrast, the mass can have uniform enhancement to that of adjacent normal

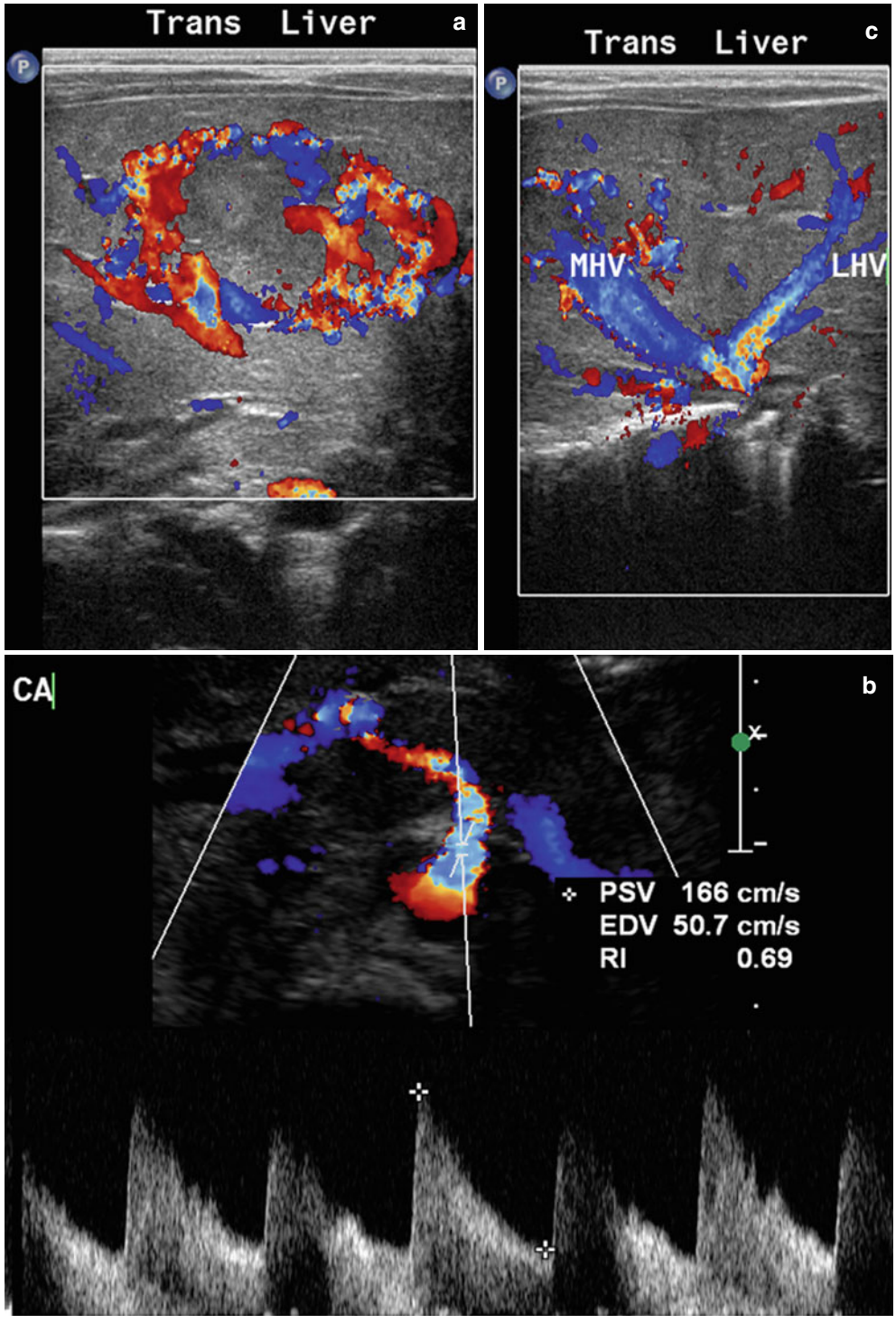


Fig. 5.1 Infantile hemangioendothelioma. Multiple ultrasound images demonstrate marked peripheral vascularity within the hemangioendothelioma (**a**). The feeding celiac artery (*CA*) is enlarged (**b**), as is the draining middle hepatic vein (*MHV*) (**c**)

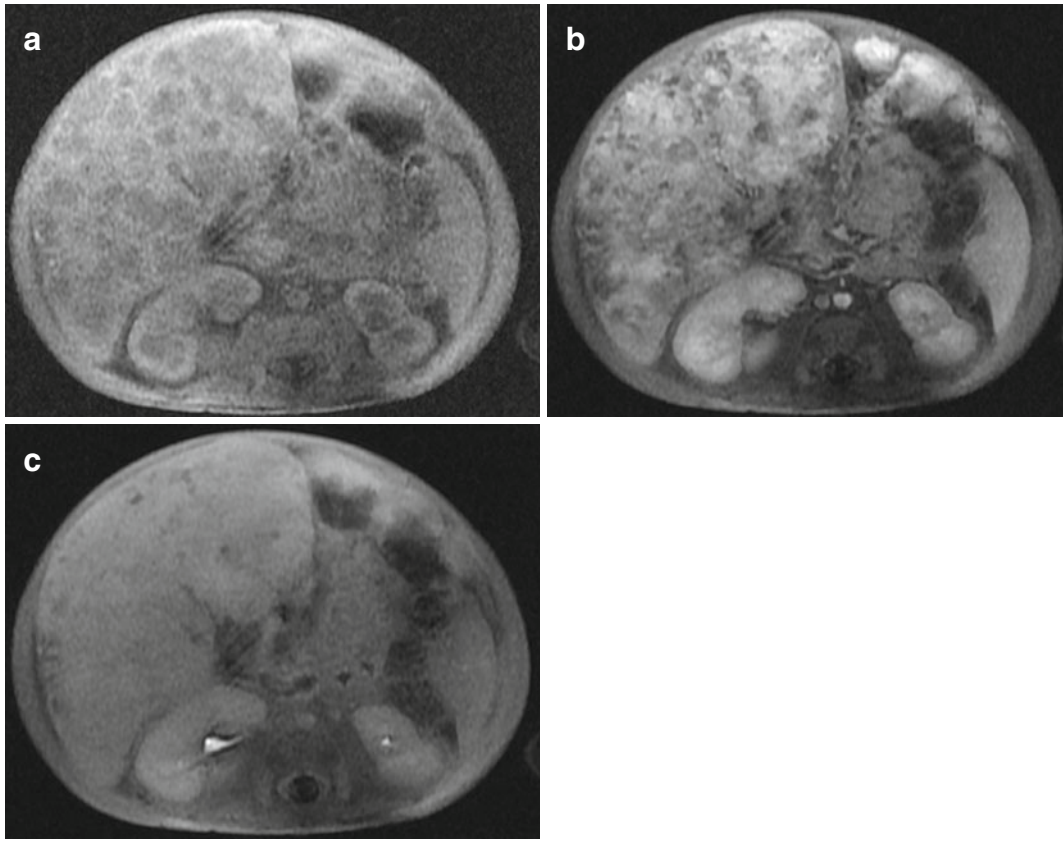


Fig. 5.2 Multiple hemangioendotheliomas on MRI. Precontrast axial T1 (**a**) demonstrates innumerable small hypointense lesions throughout the liver. Early postcontrast T1 (**b**) demonstrates heterogeneous enhancement within the liver, reflecting early peripheral enhancement. More delayed postcontrast T1 (**c**) demonstrates homogeneity of the liver, representative of progressive central enhancement of hemangioendotheliomas

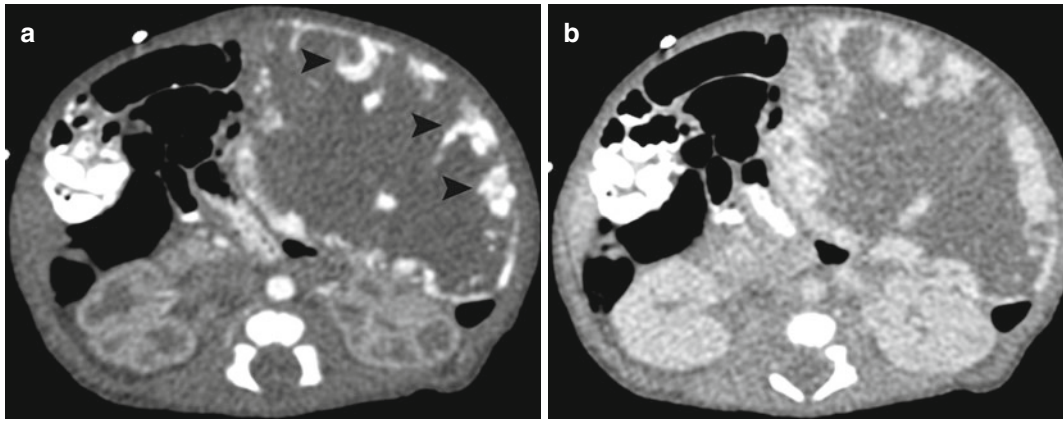
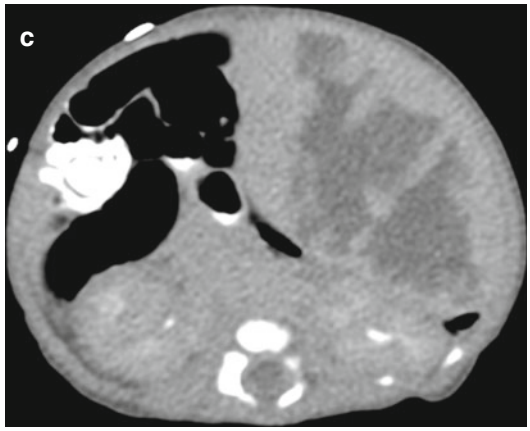


Fig. 5.3 Large hepatic hemangioma on CT. Arterial phase imaging (**a**) shows peripheral nodular discontinuous enhancement (*arrowheads*) characteristic of a hemangioma. Portal venous phase imaging (**b**) demonstrates progression of enhancement, which continues to proceed centrally as shown on delayed (excretory) phase imaging (**c**)



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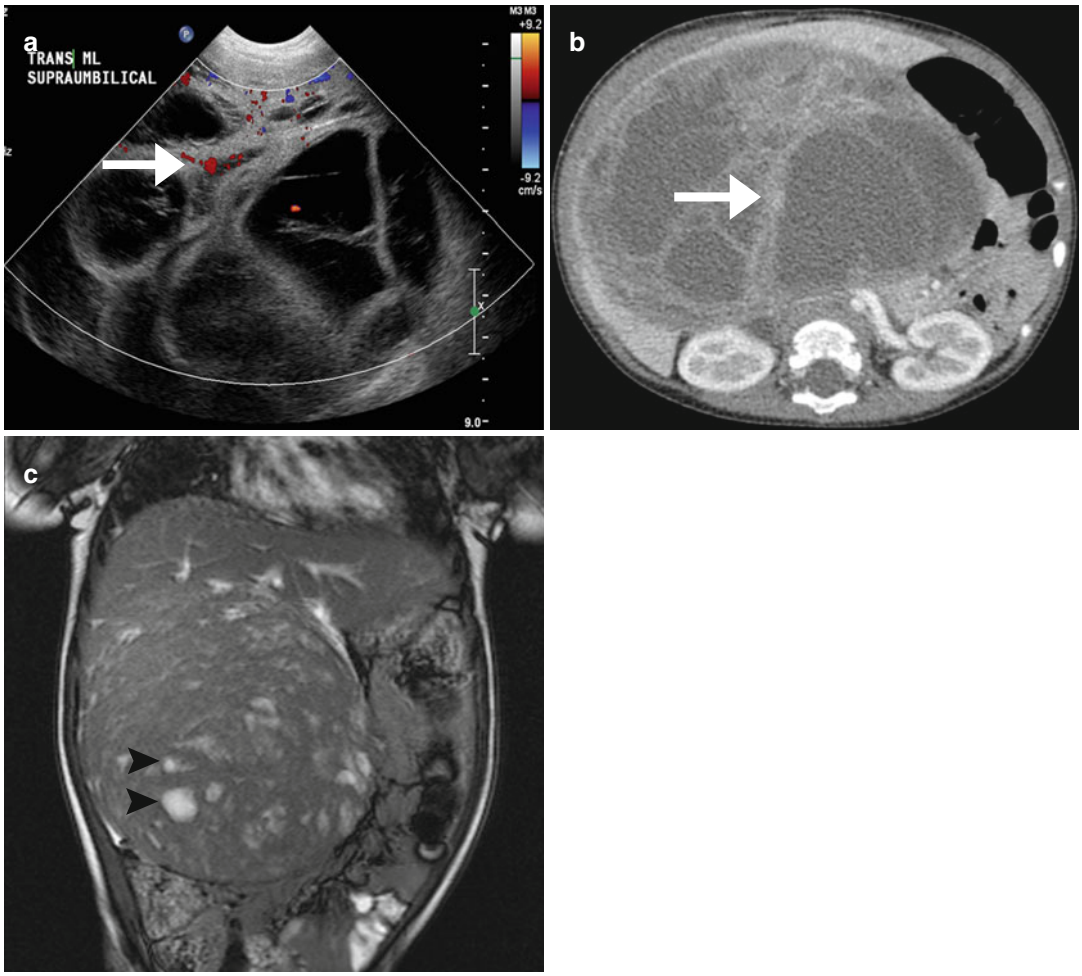


Fig. 5.4 Mesenchymal hamartoma, evaluation on multiple modalities. On ultrasound (**a**), mesenchymal hamartoma appears as a multicystic mass, with little internal vascularity except for within septations (*arrow*). The same mass was further evaluated on CT (**b**) and is seen as a well-circumscribed hypodense mass, with enhancing internal septa (*arrow*). On MRI (**c**), mesenchymal hamartoma appears as a well-circumscribed mass with internal cystic foci, which appear hyperintense (*arrowheads*) on this T2-weighted coronal image

liver and thus difficult to appreciate. The "central scar" is usually distinguishable by MRI compared to normal liver and will frequently demonstrate delayed enhancement Fig. 5.5. The vast majority of FNH will demonstrate normal uptake by nuclear scintigraphy using Technetium (Tc)-99m sulfur colloid due to the presence of normal Kupffer cells. Normal or increase uptake of sulfur colloid helps distinguish FNA from a hepatic adenoma or a malignant solid mass.

Hepatic adenoma is a benign neoplasm commonly associated with the use of oral contraceptives in young women. Most frequently it is asymptomatic but on rare occurrences can rupture leading to hemorrhage. Adenomas are typically homogenous in appearance by imaging but the presence of hemorrhagic or intracellular fat can produce some characteristic imaging features. By US adenomas can appear hypoechoic compared to adjacent liver tissue in the setting of diffuse fatty infiltration of the liver. The absence of central arterial wave pattern by Doppler US helps distinguish adenoma from FNH (which has a brisk arterial pattern from the central scar). A hepatic adenoma is typically well marginated from adjacent parenchyma by CT. Non-contrast CT will demonstrate the mass to be hypodense, hyperdense in the arterial phase, then isodense during the portal venous or delayed phase. The presence of intra-tumoral fat or hemorrhage can also produce a heterogeneous appearance Fig. 5.6. MRI of hepatic adenomas will show them to be well marginated from adjacent liver tissue and have high signal intensity on both T1- and T2-weighted imaging sequences due to the presence of fat. The enhancement pattern by MRI is similar to that of CT. Nuclear scintigraphy will demonstrate a photopenic defect by 99mTc sulfur colloid scan, but using a tracer excreted through the biliary system may show the mass to have

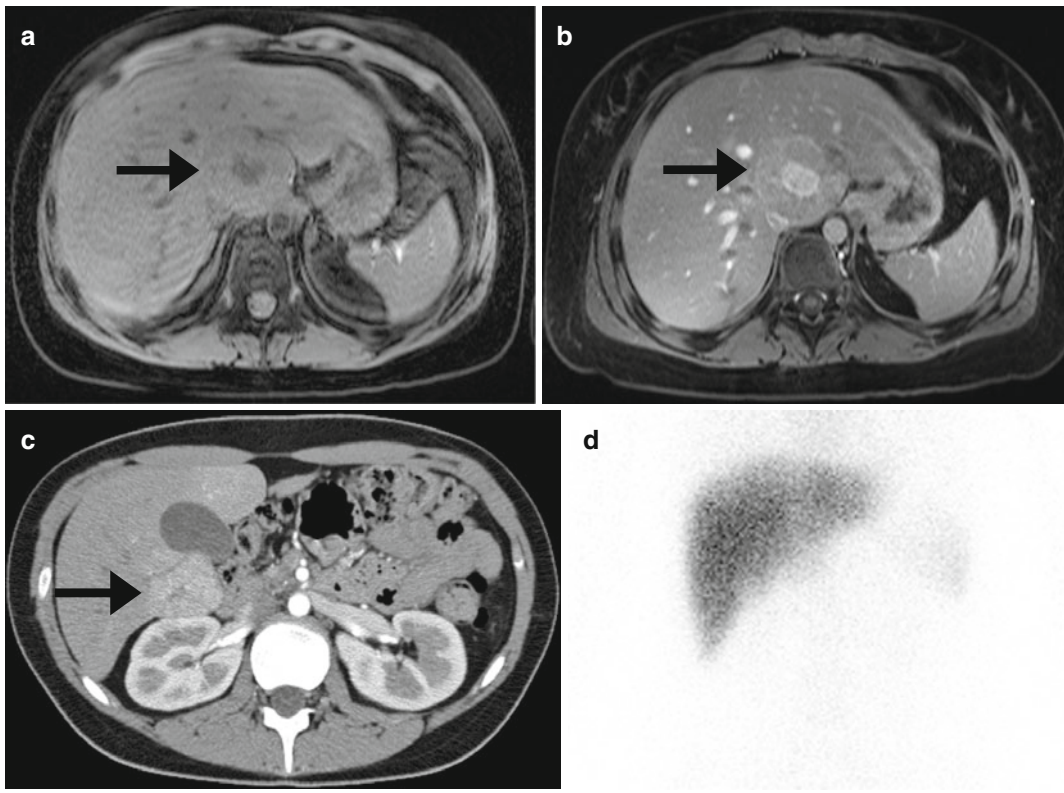


Fig. 5.5 Focal nodular hyperplasia (FNH). On early postcontrast T1-weighted MRI (**a**), FNH appears as an isointense mass with a central area of hypointensity representing the "central scar" (*arrow*). On more delayed postcontrast imaging (**b**), note that the central scar enhances, while the peripheral portion of the mass remains isointense to the liver (*arrow*). In a separate case, FNH appreciated on CT (**c**, *arrow*) demonstrates normal radiotracer uptake on a Technetium-99m sulfur colloid scan; no photopenic defect is appreciated (**d**)

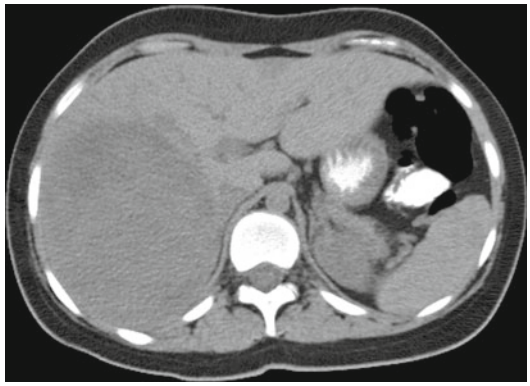


Fig. 5.6 Hepatic adenoma. Axial CT image obtained without the administration of intravenous contrast demonstrates a well-circumscribed mass which is hypodense, related to the presence of fat. In the setting of acute pain, imaging may be used to quickly assess for hemorrhage

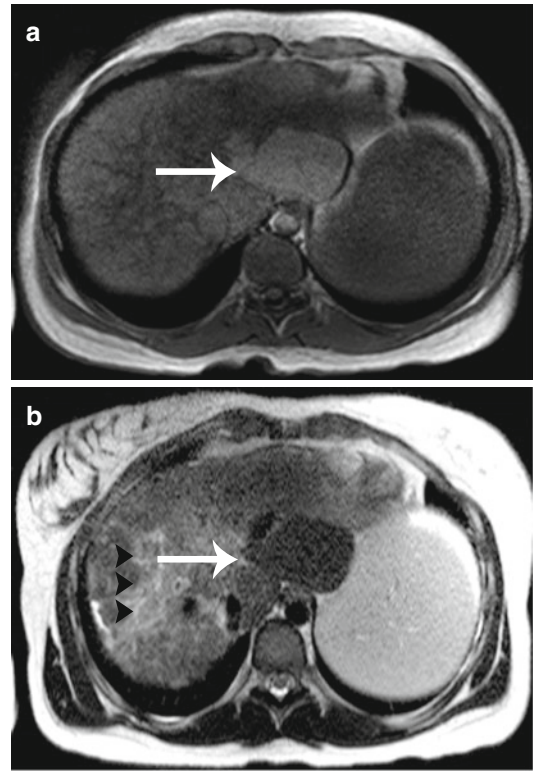


Fig. 5.7 Regenerative nodule on MRI. Axial T1-weighted imaging (**a**) obtained in a 17-year-old female with autoimmune hepatitis demonstrates a slightly hyperintense well-circumscribed mass within the posterior left hepatic lobe (*arrow*). On the corresponding T2-weighted image (**b**), the mass (*arrow*) is uniformly hypointense, compatible with a regenerative nodule. Also note the presence of T2 hyperintensity within the stroma (*arrowheads*) representing areas of fibrosis

increased radiotracer retention due to the lack of bile ducts for clearance from the mass.

Regenerating nodules can vary in size from a few millimeters to several centimeters. Given that they are composed of hepatocytes similar to surrounding parenchyma, they can be difficult to discern by imaging. Frequently there will be a nodular surface pattern of the liver indicating cirrhosis or findings related to portal hypertension can serve to suggest a history of chronic injury and regeneration. At US diffuse small regenerating nodules may simply appear as a heterogeneous liver with architectural distortion of the vascular or biliary structures. When visible by US, nodules typically will appear well circumscribed and hypoechoic. By CT the nodules are usually hypodense compared to adjacent liver on non-contrast imaging and can be isodense or hyperdense following contrast. MRI with contrast is a good imaging technique to demonstrate the extent of liver involvement and may be helpful in the assessment for malignant degeneration Fig. 5.7.

Hepatic cysts are frequent in the pediatric population. These can be seen as an incidental finding, or can be seen in the setting of polycystic disease, such as autosomal recessive polycystic kidney disease. On US, simple hepatic cysts exhibit several distinguishing characteristics, including thin walls, an anechoic internal structure, and posterior acoustic enhancement. On CT and MRI, cysts demonstrate a homogeneous appearance reflecting their water content, with no significant postcontrast enhancement. If large, hepatic cysts can cause mass effect or become symptomatic. In these circumstances percutaneous drainage and sclerotherapy can be considered Fig. 5.8.

Malignant Tumors

Similar to adults, the most common neoplasms involving the pediatric liver are related to metastatic disease associated with neuroblastoma, Wilms tumor, or lymphoma. In children, two

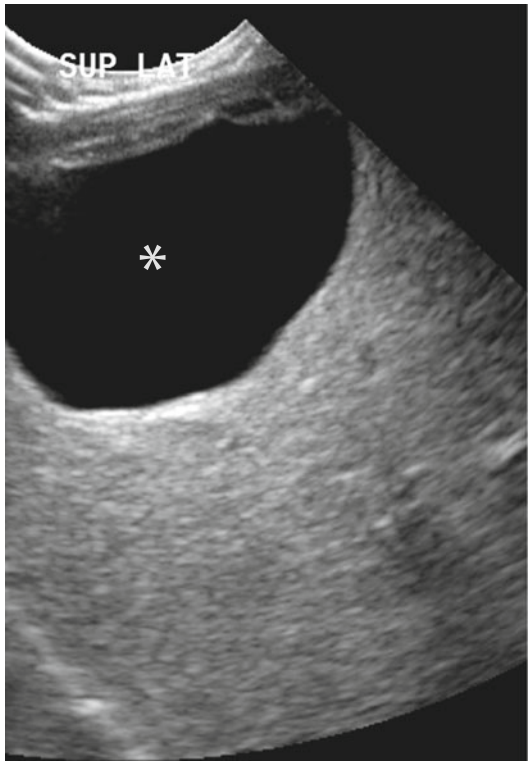


Fig. 5.8 Simple hepatic cyst on ultrasound. A lesion (*) within the right hepatic lobe meets the criteria for a simple cyst: (1) sharp, well-defined walls, (2) sonolucent, and (3) increased through sound transmission

thirds of primary liver tumors are malignant. The most frequent is hepatoblastoma followed by hepatocellular carcinoma (HCC), undifferentiated (embryonal) sarcoma, angiosarcoma, and rhabdomyosarcoma [8].

The role of imaging in assessing malignant tumors starts with defining the extent of the tumors involvement within the liver and determining if there is any extrahepatic disease [9]. US is frequently the initial screening modality to characterize the makeup of the tumor and the vascular structures of the liver. Additional cross-sectional imaging modalities (MRI and/or CT) serve as a complimentary technique to better characterize the extent of the mass, delineate anatomical borders and further assess for vascular invasion, as well as to evaluate for extrahepatic involvement.

The vast majority of *hepatoblastomas* (90 %) manifest before 5 years of age and a significant majority (68 %) present in the first 2 years. Furthermore, 80 % will present as a large mass greater than 12 cm, with the remainder as multiple hepatic masses [10]. By US most are well marginated from adjacent liver parenchyma with a heterogeneous and/or hyperechoic appearance. On CT, areas of calcification are a common finding, as these are seen in approximately 50 % of cases [10]. Following intravenous (IV) contrast for CT and MRI, hepatoblastomas will typically enhance heterogeneously and to a lesser extent than that of normal liver tissue Fig. 5.9. The critical role for imaging lies in the need to identify the extent of involvement and for the presence of vascular invasion preoperatively.

The age of presentation helps to distinguish *hepatocellular carcinoma* (*HCC*) from hepatoblastoma. HCC rarely occurs in children under 5 years of age. The US appearance is variable with typical larger lesions at the time of presentation being more heterogeneous. A hypoechoic halo around the tumor can be identified in those with a capsule [10]. Appreciating that HCC is predominantly supplied by the hepatic artery, the tumor will briskly enhance during the arterial phase by CT and MRI Fig. 5.10. During the portal venous phase, the tumor can have a similar enhancement appearance to that of normal liver or a variable appearance depending upon size, presence of intra-tumoral hemorrhage, or central tumor necrosis as a result of outgrowing arterial supply. As with hepatoblastoma, the critical role of imaging is to characterize the extent of tumor involvement within the segments of the liver, the presence of vascular invasion, and to identify extrahepatic extension. Unfortunately, due to extensive involvement of HCC, frequently the patient may not be a surgical or transplant candidate. Taking advantage of the predominantly arterial supply to the tumor, transarterial chemoembolization (TACE) for the treatment of HCC has been shown to be an effective palliative treatment option that can prolong life in adults affected with HCC [11]. Beyond palliation in children, there are descriptions of TACE serving as a treatment technique to downstage a tumor from unresectable to resectable or as a bridge until a transplant is available [12].

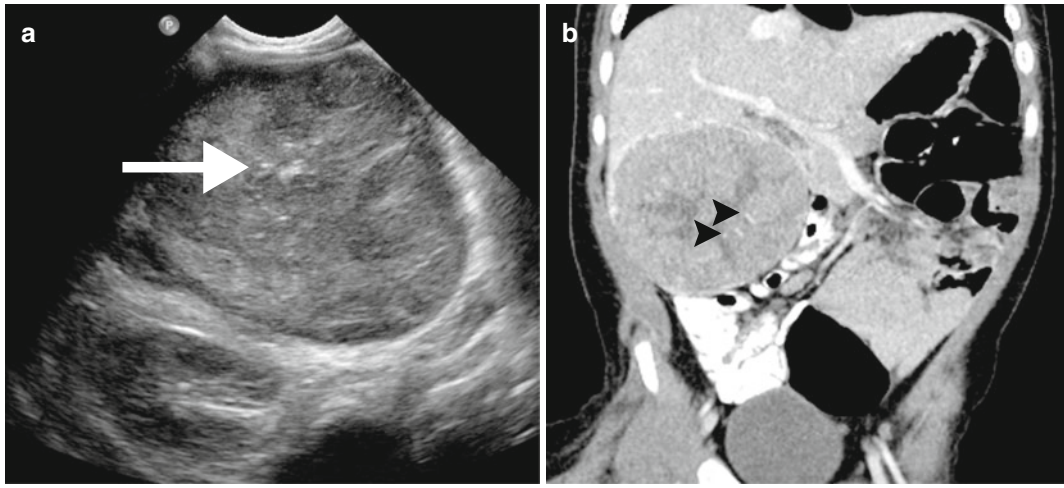


Fig. 5.9 Hepatoblastoma. On ultrasound (**a**), a heterogeneous hyperechoic mass is seen within the right hepatic lobe. Central echogenic foci (*arrow*) represent foci of calcification. Contrast-enhanced CT was subsequently performed (**b**), the coronal plane reformatted CT image similarly demonstrates a well-marginated heterogeneous mass, which enhances to a lesser degree than normal liver parenchyma. Hyperdense foci (*arrowheads*) correlate with the calcifications seen on ultrasound

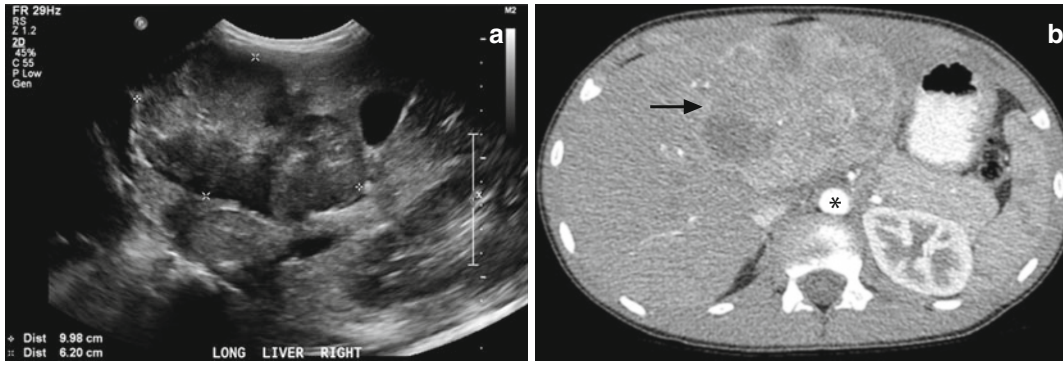


Fig. 5.10 Hepatocellular carcinoma. In this 13-year-old male, ultrasound (**a**) was performed to assess for appendicitis, but incidentally noted is a heterogeneous bilobed circumscribed mass (marked by *cursors*), with a uniform hypoechoic halo. On contrast-enhanced CT (**b**), arterial enhancement of the mass (*arrow*) is appreciated; clues indicating this is the arterial phase can be ascertained by the dense opacification of the aorta (*)

Undifferentiated sarcoma of the liver is an aggressive tumor of mesenchymal origin most frequently affecting children older than 5 years of age. The imaging features unique to undifferentiated sarcoma of the liver are a result of the myxoid component. As a result undifferentiated sarcomas appear to be a solid tumor by US but have a cystic appearance by MRI and/or CT Fig. 5.11.

Angiosarcomas and embryonal rhabdomyosarcoma are rare malignant tumor that can occur anywhere in the body including the liver in children. By imaging these tumors have nonspecific features of a heterogeneous solid liver tumor and are frequently invasive, extending beyond the margins of the liver.

Congenital Neonatal Cholestasis

Biliary atresia (*BA*) is an important cause of neonatal jaundice that must be distinguished from neonatal hepatitis to permit the opportunity for early surgical correction. It accounts for greater than 90 % of obstructive cholestasis cases [13].

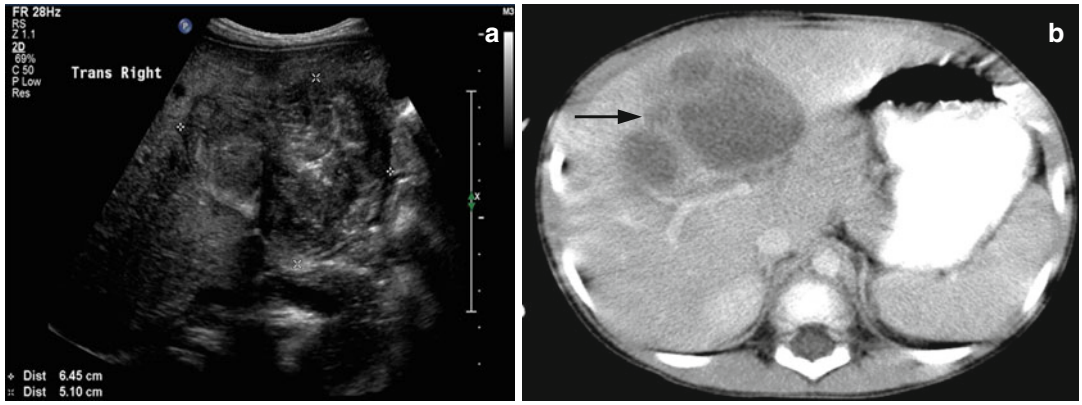


Fig. 5.11 Undifferentiated sarcoma. On ultrasound (**a**), the partially circumscribed mass (*cursors*) is heterogeneous and solid appearing. However, when evaluated on contrast-enhanced CT (**b**), the mass demonstrates low attenuation with intervening areas of density (*arrow*), suggesting it is cystic in nature

US and nuclear medicine studies are the imaging studies of choice to help differentiate these two conditions and are noninvasive. US visualization of a gallbladder favors the diagnosis of neonatal hepatitis, but 20 % of infants with BA may have a gallbladder. The *triangular cord sign* is a commonly accepted finding by US to diagnose biliary atresia [14]. It refers to an echogenic focus that represents the remnant of the obliterated biliary tract seen in the vicinity of the portal vein. Although the sensitivity of this sign is variable by reports, the specificity is regularly greater than 95 % [15]. Nuclear medicine hepatobiliary studies using a radiopharmaceutical that is excreted into the biliary system is a good physiologic imaging study to differentiate between BA and neonatal hepatitis. The excretion of the radiotracer into the bowel excludes the diagnosis of BA. Neonates with BA are able to extract the radiotracer into the liver parenchyma but fail to excrete and pass into the small bowel. An important element of the hepatobiliary scan to distinguish these two entities is that the optimal exam requires a 5-day preparatory course of phenobarbital. The purpose of this premedication is to prevent false positives. In neonatal hepatitis, liver function is impaired, and consequently, excretion into the bowel may be markedly delayed; administration of phenobarbital optimizes liver metabolism of radiotracer Fig. 5.12.

Choledochal cysts are congenital dilations of the common duct. The classic clinical presentation is an infant or young child who presents with jaundice, abdominal pain, and mass. US is the imaging technique of choice to diagnose issues of the biliary ducts and identify choledocal cysts. These will be cystic structures that are in direct communication with the biliary ducts. There are five types: *Type I* is the most common and involves dilation of the common duct Fig. 5.13. *Type II* are diverticular dilations arising off the common duct; *Type III* are choledochoceles; *Type IV* involves cystic dilation of intra- and extrahepatic biliary ducts; and *Type V* or Caroli disease involves only intrahepatic biliary ducts [16] Fig. 5.14.

Infections of the Liver

Viral hepatitis beyond the perinatal period (associated with jaundice) rarely requires imaging during the acute phase of the infection. When present any imaging modality is capable of confirming the hepatomegaly appreciated on physical exam. As a result, US should be the modality of choice for initial imaging screening. In the acute phase, no specific appearance has been described. In fact, the most common appearance is hepatomegaly [17]. Though classically the "starry sky" appearance of the liver has been described, referring to relative echogenicity of the portal triads relative to diffusely edematous liver parenchyma, this has not proven to be a sensitive finding [18]

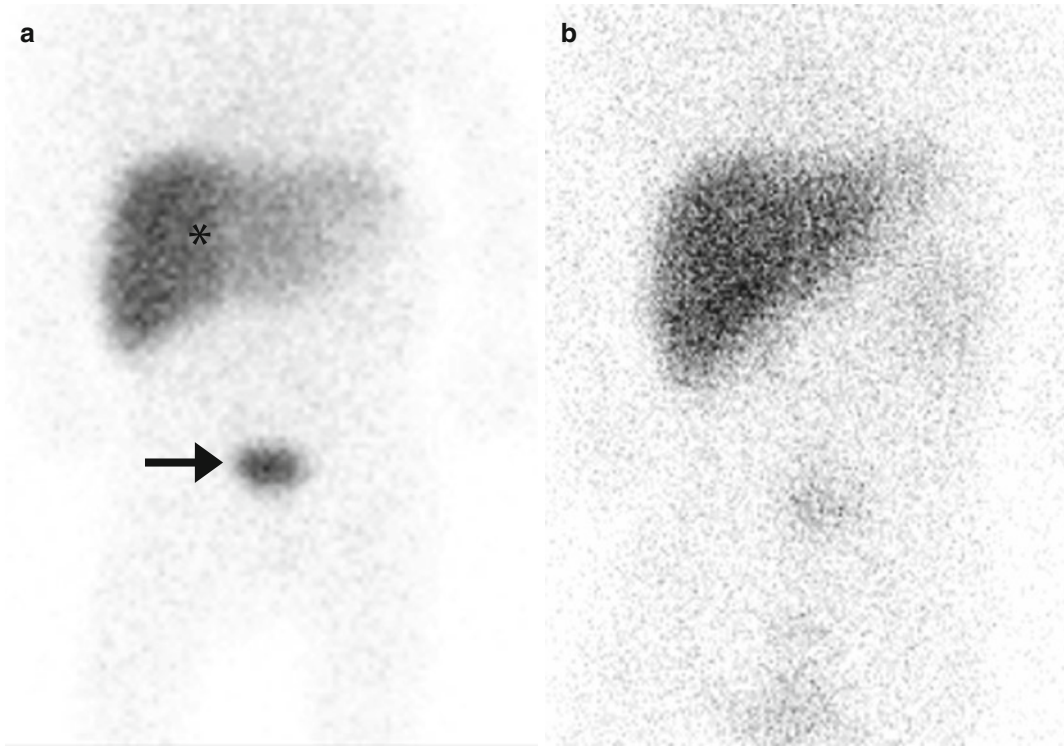


Fig. 5.12 Biliary atresia evaluation using hepatobiliary scan. (**a**) Initial planar image on the left obtained at 1 h post radiotracer injection demonstrates activity within the liver (*) as well as within the urinary bladder (*arrow*). (**b**) Planar image on the right obtained at 24 h demonstrates diffuse hepatic activity without evidence of bowel uptake, compatible with biliary atresia

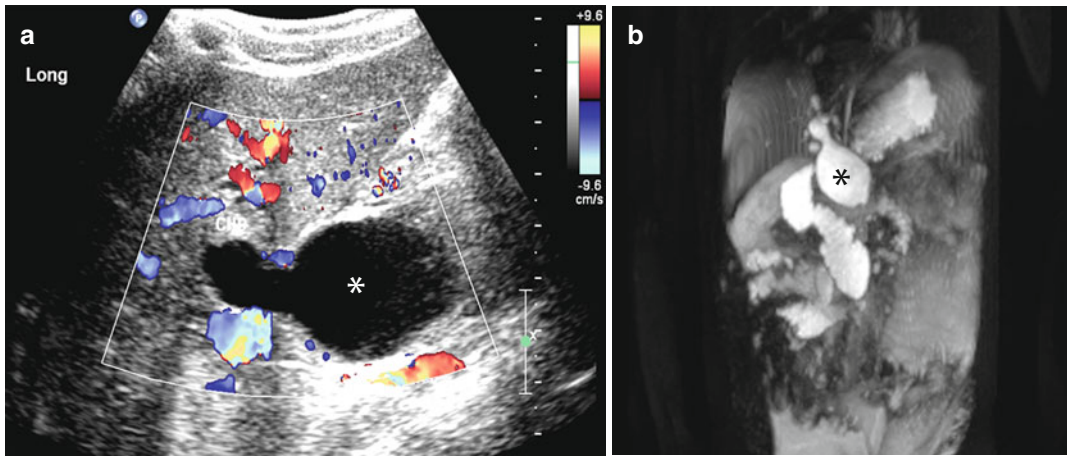
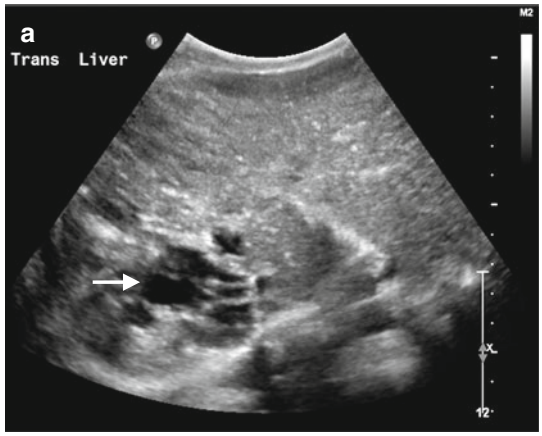


Fig. 5.13 Choledochal cyst, type I. Ultrasound image (**a**) demonstrates fusiform dilatation of the common bile duct (*). Color flow confirms that this is indeed a biliary structure. Coronal maximal intensity projection (MIP) reconstruction from a magnetic resonance cholangiopancreatogram (MRCP) (**b**) confirms this finding of a dilated common bile duct (*) and better demonstrates the anatomy



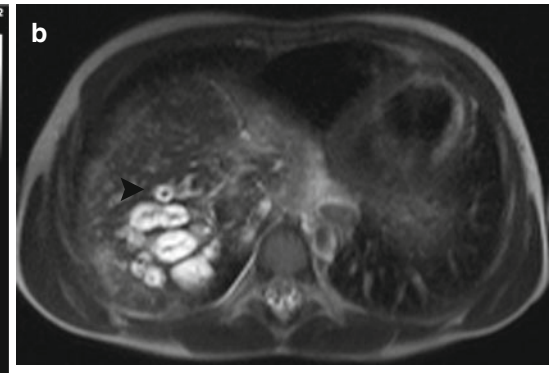


Fig. 5.14 Caroli disease (choledochal cyst, type V). On initial ultrasound evaluation (**a**), cystic foci are focally identified within the posterior right hepatic lobe (*arrow*). Subsequently MRI was performed (**b**) and on axial T2-weighted imaging, multiple dilated biliary ducts are identified in the posterior right hepatic lobe as areas of hyperintensity. Note the presence of central hypointensity (*arrowhead*), which represents the normally sized portal radicle

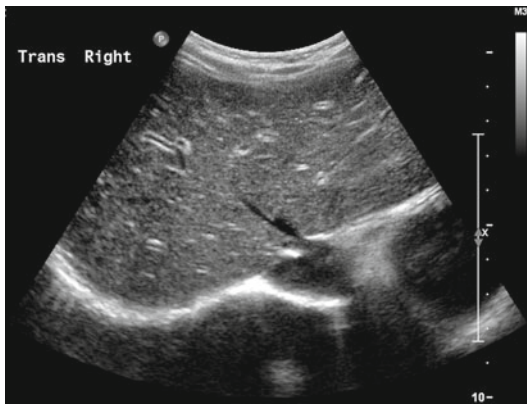


Fig. 5.15 Viral hepatitis related to hepatitis B. There is diffusely decreased echogenicity of the liver parenchyma, relating to diffuse edema, with echogenic foci representing normal portal triads, which are accentuated in this setting. This appearance, the so-called "starry sky" pattern, is not specific to viral hepatitis and can also be seen in heart failure or infiltrating malignancy

Fig. 5.15. In the chronic phase, changes of cirrhosis may develop. Gallbladder wall thickening can also be present as a result of edema.

Pyogenic abscesses are typically associated to children that are immunocompromised or immunosuppressed. Other children susceptible to the development of pyogenic abscess include those with chronic granulomatous diseases, inflammatory bowel disease, or those with other intra-abdominal infections. By US the dominant abscess will be predominantly hypoechoic internally and surrounded by a hypoechoic halo representing hepatic parenchyma edema. There is frequently debris within the abscess, and thus the collection is not simply an anechoic structure as would be a simple cyst. Contrast-enhanced CT and MRI will demonstrate enhancement of the abscess wall frequently surrounded by a halo of parenchymal that has less enhancement compared to normal liver as a result of edema Fig. 5.16. Internally the abscess will have little internal enhancement except for some internal septations, which helps differentiate these from solid tumors. Satellite microabscesses may also be present, which may be identified on cross-sectional imaging: on CT and MRI, these appear as areas of decreased enhancement, while on US, these demonstrate a hypoechoic appearance. There should not be uptake of radiotracer in the lesion by nuclear scintigraphy.

Fungal and parasitic infections can also affect the liver of children. Findings of fungal infections are typically nonspecific and when multiple small lesions are present will appear as hypoechoic lesions by US and hypodense following IV contrast by CT or MRI. Amebic abscesses appear similar to pyogenic abscesses. Echinococcal cysts often exhibit a dominant cyst with several smaller "daughter" cysts Fig. 5.17.

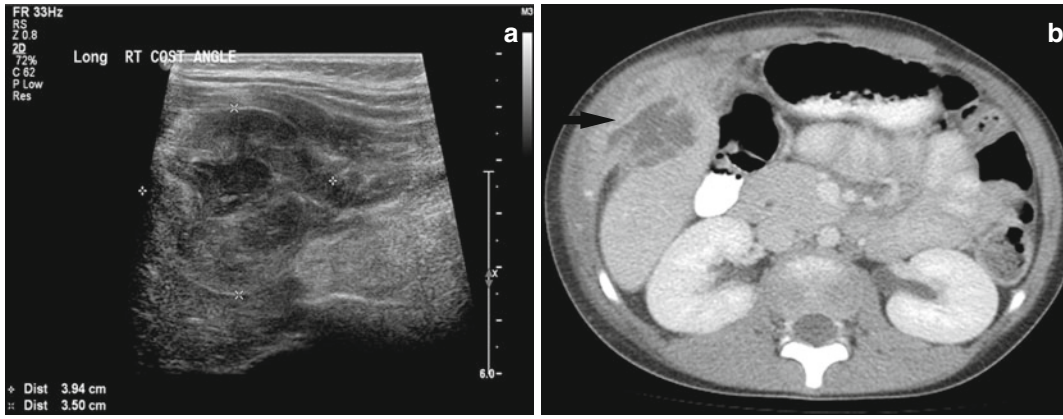


Fig. 5.16 Pyogenic abscess. On initial ultrasound evaluation for right upper quadrant pain (**a**), a heterogeneous mass is appreciated (*cursors*) with no appreciable internal vascularity (color flow image not shown). Abscess was clinically suspected and CT subsequently performed (**b**). On contrast-enhanced CT, a peripherally enhancing fluid collection extends beyond the liver capsule into the overlying anterior abdominal wall (*arrow*). Aspirate yielded methicillin-sensitive *Staphylococcus aureus*

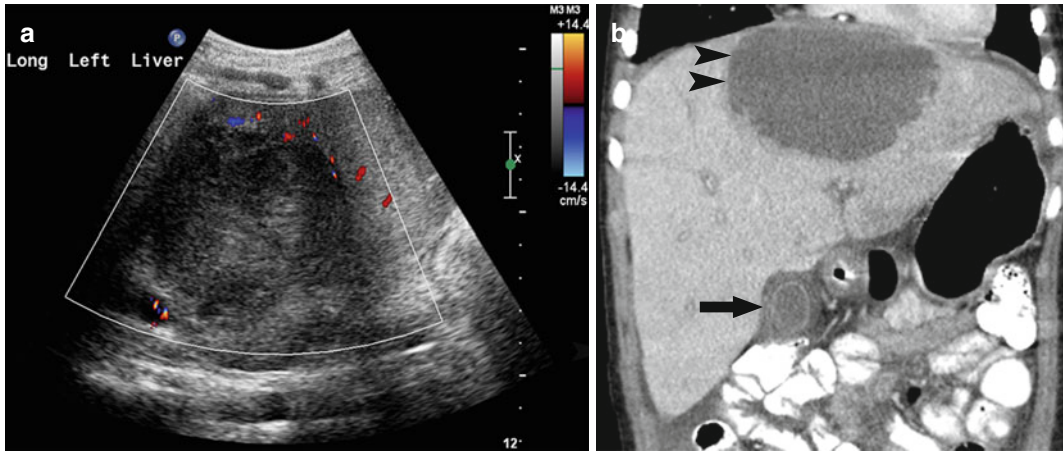


Fig. 5.17 Parasitic abscess. Ultrasound evaluation (**a**) in a child with right upper quadrant pain and a recent travel history to Mexico demonstrates a heterogeneous collection with peripheral vascularity, suspicious for abscess. Contrast-enhanced CT (**b**) performed to better define the extent of the collection demonstrates a low density collection with peripheral enhancement (*arrowheads*). Aspirate yielded *Entamoeba histolytica*. Note also the presence of gallbladder wall thickening (*arrow*), a nonspecific finding

Diffuse Hepatic Parenchymal Disease

Cirrhosis is a chronic condition that can be a result of numerous diseases (congenital or acquired) in children resulting in diffuse fibrotic replacement of the normal hepatic parenchyma. The key role of imaging is to confirm the clinical diagnosis, provide a means to follow the disease progression or response to therapy, and evaluate for complications associated with cirrhosis such as portal hypertension or the development of malignancies. Imaging features include a nodular surface pattern of the liver with heterogeneous appearance of the parenchyma. Typically the right lobe is small with compensatory hypertrophy of the caudate lobe and even the lateral segment of the left lobe. US is a good initial imaging study and can demonstrate diffuse increased echogenicity of the parenchyma Fig. 5.18. Contrast-enhanced CT and MRI can demonstrate

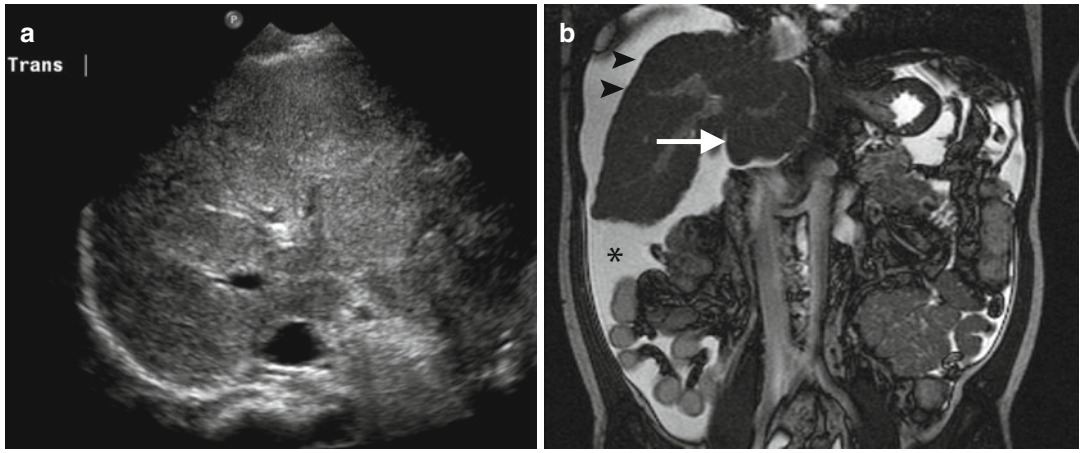


Fig. 5.18 Cirrhosis. On sonographic evaluation (**a**), there is marked heterogeneity of the liver parenchyma. T2-weighted coronal MRI (**b**) demonstrates a shrunken nodular contour to the right hepatic lobe (*arrowheads*). Moderate amount of ascites is appreciated (*). Finally, note compensatory hypertrophy of the caudate lobe (*arrow*)

the diffuse heterogeneity due to regenerating nodules in the background of fibrosis. It can be difficult to differentiate a regenerating nodule from hepatocellular carcinoma by any imaging modality. However, MRI may be more sensitive in distinguishing regenerating nodule versus tumor using various imaging sequences before and after contrast administration, as previously shown (Fig. 5.7).

Fatty infiltration of the liver can be seen in diseases beyond that of cirrhosis. US will show diffuse increased echogenicity of the liver, CT will demonstrate diffuse hypodensity, and MRI will reveal signal intensity on the various sequences that correspond to fat (such as fat suppression sequences) Figs. 5.19 and 5.20. With *hemochromatosis*, by contrast, excessive iron deposition results in diffusely increased hepatic density on CT and strikingly decreased signal intensity on MRI. US is often unrevealing in children with hemochromatosis.

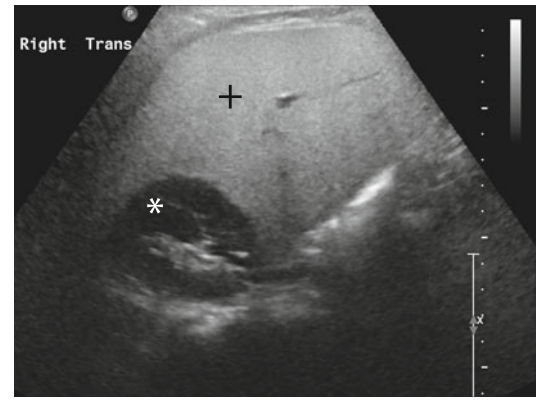


Fig. 5.19 Fatty infiltration of the liver, ultrasound evaluation. On this ultrasound image of the right hepatic lobe, the liver (+) demonstrates a markedly echogenic appearance relative to the right kidney (*). Normally, the liver is isoechoic to slightly echogenic relative to the kidney

Portal Hypertension

Imaging is critical in determining the anatomic level of vascular obstruction and can have great implications regarding the treatment options available for children with complications associated with portal hypertension. Although frequently associated with cirrhosis, *portal hypertension* can also occur as a result of extraparenchymal vascular conditions. Classically portal hypertension can be classified by the anatomic level of portal flow resistance: prehepatic, intrahepatic, or posthepatic [19]. Secondary imaging findings to support portal hypertension can include evidence of cirrhosis, ascites, splenomegaly, varices, an enlarged main portal vein, and a hepatofugal flow pattern Fig. 5.21. *Congenital arterial portal fistulas* can also result in neonatal portal hypertension due to high inflow.

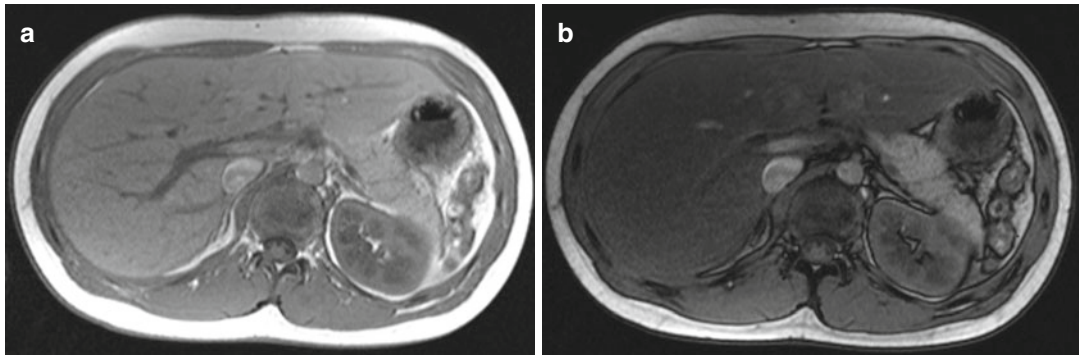


Fig. 5.20 Fatty infiltration of the liver, MRI. (**a**) Chemical shift imaging is used to demonstrate the presence of intracellular fat; (**b**) signal dropout within the liver on "out-of-phase" imaging, as seen on the image at right, reflects such fat deposition

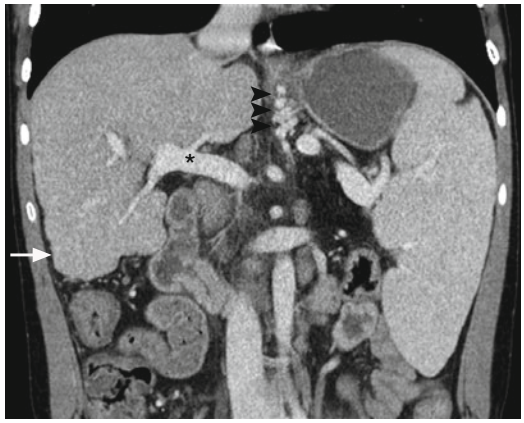


Fig. 5.21 Portal hypertension, secondary findings. On this coronal contrast-enhanced CT, note nodularity of the liver (*arrow*) seen in the setting of cirrhosis. Correspondingly, the main portal vein is enlarged (*), gastroesophageal varices are present (*arrowheads*), and marked splenomegaly is noted

Doppler and spectral waveform US offers a noninvasive method to assess and characterize in real time for hepatofugal flow, patency of the portal vein, varices, or for abnormal fistulous communications. MRI and CT are useful to give a global anatomical assessment of the liver parenchyma and hepatic vasculature, to clarify etiology of the portal hypertension, and assess patency of hepatic vascular structures including the portal vein, for planning of a surgical shunt or a transjugular intrahepatic portosystemic shunts (TIPS). TIPS, which will be discussed in detail later, is a percutaneous endovascular image-guided procedure that has been shown to be effective in treating variceal bleeding associated with portal hypertension in children and infants [20].

The pediatric interventional radiologist can be called upon to perform a percutaneous or transjugular route catheter portal venographic study to directly characterize portal flow, measure portal pressure to determine the portal systemic gradient, demonstrate, and embolize or sclerose varices.

US is a good screening tool for the evaluation of extrahepatic portal vein obstruction as the etiology for *prehepatic portal hypertension*. Chronic main portal vein occlusion will frequently demonstrate an extensive collateral network or *cavernous transformation* in the absence of an identifiable main portal vein by contrast-enhanced CT or MRI Fig. 5.22. Catheter arteriogram of the superior mesenteric artery with delayed angiographic imaging into the portal venous phase is useful to demonstrate the flow pattern of the portal supply and the patency of the intrahepatic portal structures. Determining the patency and location of the left intrahepatic portal vein is valuable in order to determine the feasibility for the construction of a surgical Rex shunt to relieve the portal hypertension. The Rex shunt joins the extrahepatic portal vein to the umbilical segment of the intrahepatic left portal vein [21].

Acute portal vein thrombosis is well demonstrated with US. Color Doppler US will show absence of flow in the portal vein, and grayscale

imaging can show the portal vein to be hyperechoic and even distended due to the clot. Contrast-enhanced MRI and CT may be required to determine if the mesenteric veins or splenic veins are also involved [22] Fig. 5.23.

Budd-Chiari syndrome resulting from obstruction of the hepatic veins or suprahepatic IVC can result in *posthepatic portal hypertension*. The outflow obstruction acutely can lead to hepatic enlargement and a heterogeneous echotexture by US. The major hepatic veins may not be identified by US. Contrast-enhanced CT and MRI will demonstrate an enlarged heterogeneous liver and the absence of identifiable major hepatic veins. There is also prolonged retention of contrast. Caudate lobe hypertrophy is present in 75 % of patients due to the separate venous drainage into the IVC [22].

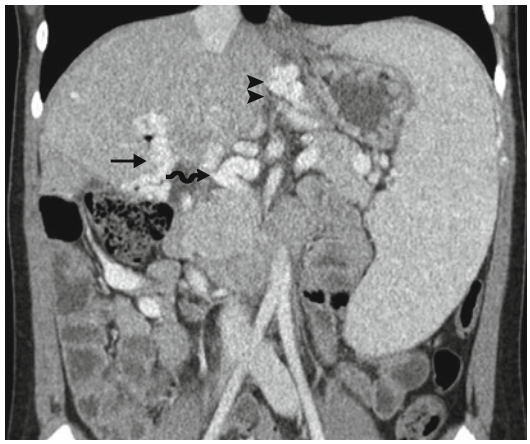


Fig. 5.22 Portal hypertension associated with cavernous transformation as a result of main portal vein occlusion. On this coronal contrast-enhanced CT, the main portal vein is not identified, and intrahepatic cavernous transformation is seen (*arrow*). Associated secondary findings of portal hypertension, including gastroesophageal varices (*arrowheads*) supplied by an engorged coronary vein (*curved arrow*), as well as marked splenomegaly is present

Liver Transplantation Imaging

Following an orthotopic liver transplant in a child, the clinical presentation of acute rejection is nonspecific and the imaging findings are often unrevealing. Therefore, a biopsy is required to diagnose acute rejection. The major role of imaging is to exclude complications that are potentially correctable. Imaging of post transplant complications can be separated into three major categories: (a) vascular complications, (b) biliary tract complications, and (c) perihepatic fluid collections [23]. Furthermore, imaging also offers the opportunity to potentially serve as a tool to perform percutaneous minimally invasive treatment that addresses the various potential complications.

Doppler sonography is the primary initial noninvasive imaging modality to screen for

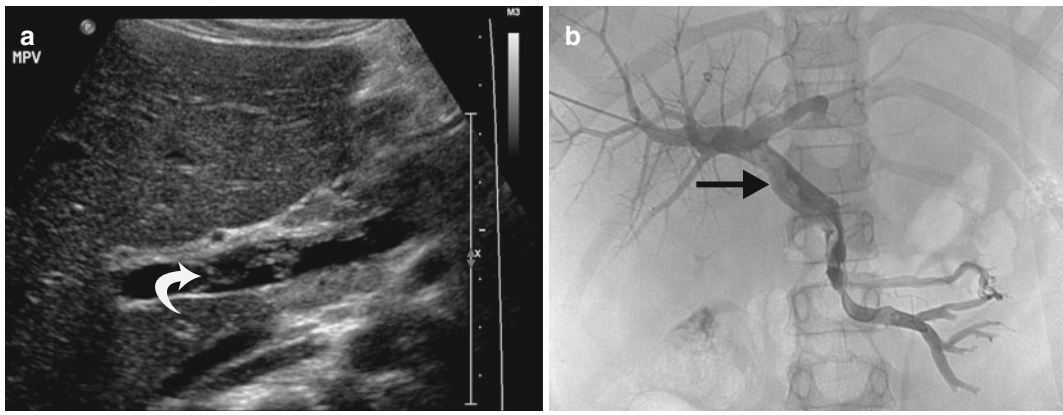


Fig. 5.23 Portal vein thrombosis. On grayscale ultrasound (**a**), note the presence of echogenic debris (*curved arrow*) within the main portal vein, representing thrombus. The patient was then referred for catheter-directed thrombolysis. (**b**) A transhepatic portal venogram performed prior to thrombolysis demonstrates the corresponding filling defect (*arrow*) representing thrombus within the main portal vein

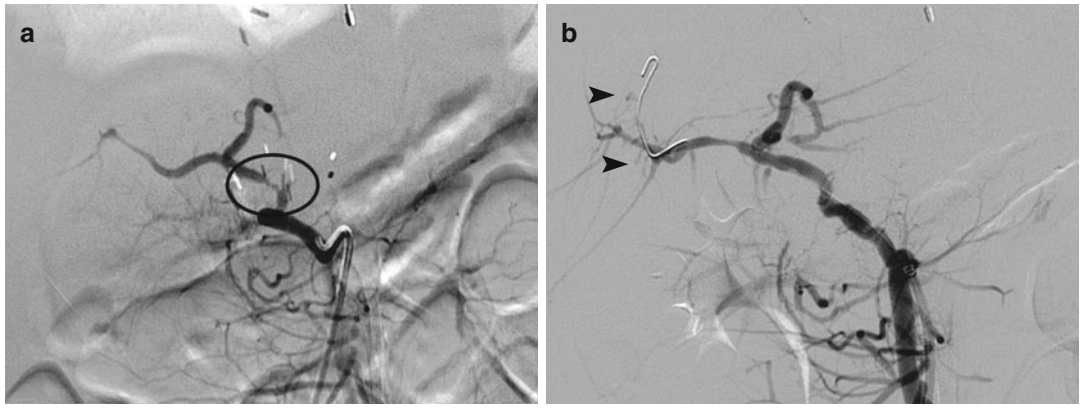


Fig. 5.24 Hepatic artery stenosis after liver transplant. Digitally subtracted angiographic image obtained in an 8-month-old child post transplant initially demonstrates high-grade stenosis of the hepatic artery (**a**, *circled*) with poor hepatic perfusion. Following balloon angioplasty, repeat angiogram was performed (**b**) and demonstrates improvement in caliber of the hepatic artery. Liver perfusion has improved, as signified by increased arborization of arterial vessels (*arrowheads*)

vascular complications. The most common vascular complication involves the hepatic artery followed by the portal vein. Less frequently, hepatic vein or IVC complications can be encountered. Hepatic artery thrombosis is identified as the absence of flow while hepatic artery stenosis will demonstrate increased velocity at the point of stenosis with associated diminished resistive index (RI) and a parvus-tardus waveform beyond the stenosis. An RI of less than 0.50 and a prolonged systolic acceleration time greater than 0.08 s is highly suggestive of a thrombosis or high-grade stenosis. In the acute period following transplant, surgical revision is often necessary. However, beyond 2 weeks it is not uncommon for an interventional radiologist to perform catheter-directed thrombolysis or angioplasty +/− stenting of the culprit vessel Fig. 5.24.

Similarly, US can be used to screen for portal vein or hepatic vein stenosis. CT and MRI although capable of identifying an anatomical narrowing cannot determine the hemodynamic significance of a narrowing. If an abnormality is identified by noninvasive imaging, then proceeding with a percutaneous catheter study for confirmation would be warranted. Although invasive, percutaneous interventions have several benefits. It can definitively confirm the diagnosis with anatomic and physiologic pressure gradient testing as well as offer the opportunity to treat a thrombosis or stenosis with angioplasty or stenting during the same session.

When biliary tract complications, such as bile duct stenosis/stricture with resultant obstruction, are clinically suspected, it needs to be aggressively evaluated. US is a good initial screening method and can demonstrate the dilated biliary ducts. However, it is not uncommon that despite a severe stenosis and clinical evidence for obstruction, biliary ductal dilation will not be present or identified by noninvasive imaging. Anastomotic strictures are frequently a result of fibrosis, whereas intrahepatic strictures are often associated with a history of hepatic arterial compromise leading to biliary ischemia [23]. As a result a *percutaneous transhepatic cholangiogram* (*PTC*) can be performed to directly visualize the biliary system and identify point(s) of stenosis Fig. 5.25. This also offers the opportunity to dilate the stricture(s) and position an internal-external biliary drain for decompression of the obstructed system. CT or MRI is useful to determine if there is an extrinsic compressing causing obstruction of the duct.

The final category in which imaging plays a primary role is for the identification of infected perihepatic fluid collections. These can arise as a

result of a bile leak or the development of an abscess (bacterial or fungal). Once again US is a good screening tool, but frequently CT or MRI are necessary to provide information regarding the full extent of involvement and to help determine if image-guided percutaneous drainage tube placement is feasible. Nuclear scintigraphy can be used to confirm that a fluid collection is a result of a bile leak Fig. 5.26.

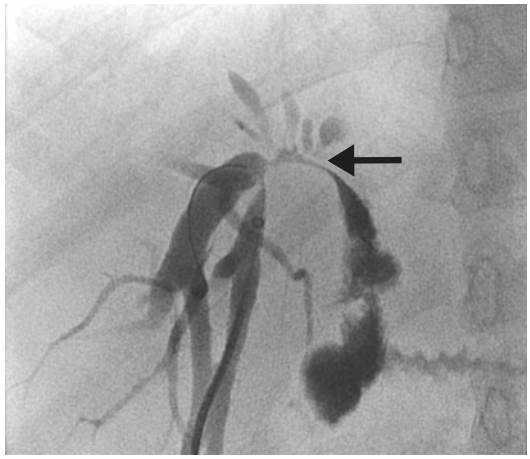


Fig. 5.25 Biliary stricture. Percutaneous transhepatic cholangiogram demonstrates a high-grade anastomotic stricture (*arrow*) in an 8-year-old girl with a split liver transplant

Image-Guided Hepatic Interventions

The utility of imaging has evolved beyond that merely of a diagnostic method in the evaluation of a child's liver. It can also serve as a tool to guide and direct a pediatric interventional radiologist during the performance for the successful execution of a wide variety of minimally invasive percutaneous procedures with either further diagnostic or even therapeutic intent.

In general percutaneous liver biopsies do not require image guidance. However, in the setting of uncorrectable coagulopathy and/or the need to maintain anticoagulation or antiplatelet therapy, a *transjugular liver biopsy* is a reliable alternative image-guided technique that can be used even in children with liver transplants [24]. In addition image guidance is useful when a discrete hepatic lesion needs to be directly sampled. US has the benefit of permitting real-time guidance as well as permitting more diverse off-angle capabilities during the performance of a directed *percutaneous liver biopsy*. US also is the imaging modality of choice for the positioning of a percutaneous *radiofrequency ablation* (*RFA*) probe in the treatment of children with discrete hepatic masses. RFA and other local regional

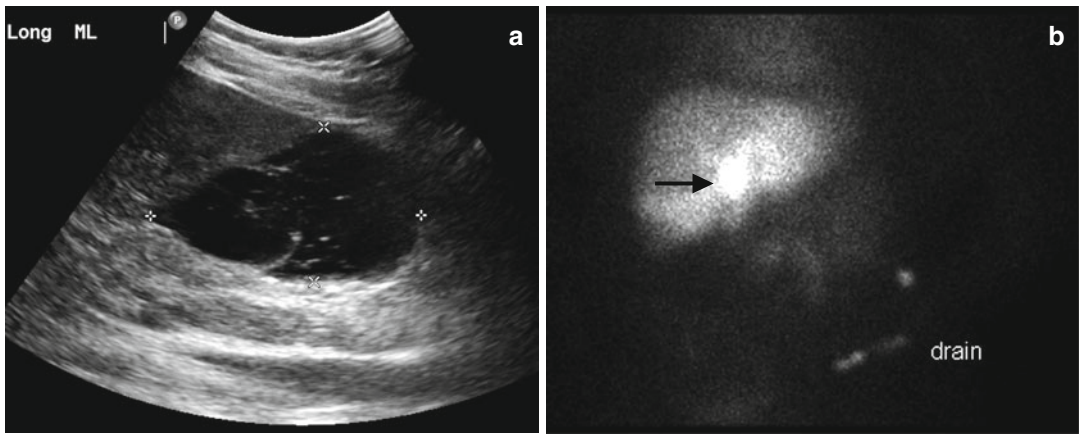


Fig. 5.26 Bile leak, status post transplant. On ultrasound (**a**), a perihepatic fluid collection was identified on initial postoperative imaging (*cursors*). Due to increasing size, bile leak was suspected, and hepatobiliary scan was performed (**b**). On 4 h delayed image, focal radiotracer activity was demonstrated in the right upper quadrant (*arrow*). In a patient status post cholecystectomy, this is representative of bile leak. Note also the presence of radiotracer at the level of an external drain

techniques such as *transarterial chemoembolization* (*TACE*) are increasingly becoming available to pediatric patients as a minimally invasive option for the treatment of localized malignant hepatic tumors. These local regional treatments can serve as an adjunct and/or potential bridge to transplant [25]. TACE takes advantage of the dual vascular supply supporting the liver, in which the majority of tumor supply is contributed from the hepatic arterial system. The hepatic artery can be selectively catheterized from a percutaneous femoral arterial access route to permit direct infusion of the chemotherapeutic agent directly into the arterial supply of the tumor while further increasing the dwell time of the agent by occluding the arterial supply with an embolic particle upon completion. This also leads to tumor devascularization. Frequently both TACE and RFA are performed to address a hepatic tumor and can be complementary in order to treat the entirety of a localized tumor.

Percutaneous *transjugular intrahepatic portal systemic shunt* (*TIPS*) procedure is a feasible alternative to surgical shunt procedures in children with medically uncontrollable complications associated with portal hypertension such as GI bleeding, refractory ascites, hepatic hydrothorax, or hepatorenal syndrome. The construction of a TIPS consists of deploying a stent (frequently a covered endograft) across the hepatic parenchymal tract to join the hepatic venous outflow directly to the portal venous inflow. This effectively forms a shunt that serves to relieve the portal hypertension and decompress varices. TIPS in children and infants has been shown to be clinically effective and durable with low complication rates and do not preclude the possibility to perform a liver transplant in the future [20] Fig. 5.27.

A transjugular approach similar to TIPS or a percutaneous transhepatic approach using US can be performed to gain direct access into an acutely thrombosed portal vein. Spontaneous clearance of acute portal vein thrombosis is unlikely [26]. *Catheter-directed thrombolysis* (*CDT*) permits the ability to directly infuse a thrombolytic agent via a catheter embedded into the clot. A thrombectomy procedure can be performed during the same session as CDT allowing for a lower dose requirement of a thrombolytic agent and speeds up time to achieve clearance compared to systemic intravenous infusion or indirect mesenteric arterial catheter infusion when addressing acute portal vein thrombosis [27].

Percutaneous image-guided *abscess drainage tube* placement procedures are well suited to address hepatic abscesses. US guidance is preferred in children to reduce the need to use ionizing radiation, but CT imaging may be required depending upon abscess location. The placement

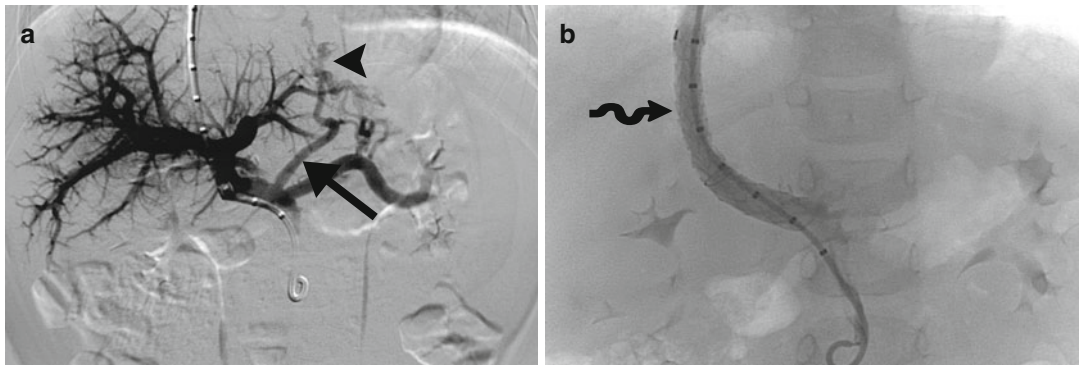


Fig. 5.27 Transjugular intrahepatic portosystemic shunt (TIPS). 17-month-old male with biliary atresia and portal hypertension presenting with recurrent upper GI bleeding. Digitally subtracted image of a portal venogram (**a**) demonstrates opacification of not only the main portal vein but also the coronary vein (*arrow*) and esophageal varices (*arrowhead*). TIPS was constructed, and follow-up (**b**) demonstrates contrast passage through the shunt (*curved arrow*); the coronary vein and varices are no longer opacified. Portal systemic gradient decreased from 14 mmHg pre-TIPS to 5 mmHg post-TIPS

of a drainage tube to drain the abscess is more effective than needle aspiration alone [28]. Percutaneous drainage has been shown to be effective and safe even in the presence of complex and multiple hepatic abscesses [29].

Complications of liver transplants that may benefit from image-guided intervention can be separated into three major categories as discussed under the section of transplant imaging. The needs of these children are best served in a multidisciplinary format with involvement of the pediatric transplant hepatologist, transplant surgeon, and the interventional radiologist.

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