Liver Transplantation in Children: Techniques and What the Surgeon Wants to Know From Imaging

Jaimie Nathan, MD

1. Which of the following is the most common cause of neonatal cholestasis and is the underlying diagnosis in over 50% of pediatric liver transplant recipients?
   A. Progressive familial intrahepatic cholestasis type I
   B. Biliary atresia
   C. Alagille syndrome
   D. α1-antitrypsin deficiency

Correct Answer: B

Reference:


2. Which of the following is the most common cause of postoperative morbidity in the pediatric liver transplant recipient?
   A. Hepatic venous outflow obstruction
   B. Hepatic artery thrombosis
   C. Portal vein thrombosis
   D. Biliary complications

Correct Answer: D

Reference:


Role of MRI in Pediatric Liver Transplantation (Pre- and Post-)

Geetika Khanna, MD

3. Biliary strictures are caused by compromise of which of the following structures in the transplanted liver?
   A. Hepatic artery
   B. Hepatic vein
   C. Portal vein
   D. Hepatic sinusoids
Correct Answer: A

Rationale:
Although the transplanted liver has a dual blood supply, the bile ducts are exclusively supplied by the hepatic artery. Hepatic artery stenosis/thrombosis can cause biliary ischemia, hepatic necrosis, bile leaks/bilomas and biliary strictures.

Reference:

4. Which of the following is the MOST common indication for liver transplantation in the 0-18 year age group in North America?
   A. Acute liver failure
   B. Liver malignancies
   C. Metabolic liver disease
   D. Biliary atresia

Correct Answer: D

Rationale:
The most common indications for liver transplantation in children in North America are: biliary atresia, metabolic conditions, acute liver failure, cirrhosis and liver tumors in the order listed. At least 80% of patients with biliary atresia receive a transplant by 20 years of age, with the majority needing a transplant by 5 years of age.

Reference:

Imaging of Liver Fibrosis/MR Elastography
Jonathan Dillman, MD, MSc

5. MRI assessment of liver fibrosis based on morphology and conventional signal characteristics is:
   A. Sensitive but not specific
   B. Specific but not sensitive
   C. Both sensitive and specific
   D. Neither sensitive nor specific

Correct Answer: B.
Rationale:
MRI assessment of liver fibrosis based on morphology and conventional signal characteristics is specific but insensitive. Changes in morphology and signal intensity are usually only observed when fibrosis is moderate to severe. Early/mild liver fibrosis typically does not result in visible changes in liver morphology or signal intensity.

Reference:

6. When performing MR elastography in patients with hepatic iron deposition, what is the best scanner field strength and pulse sequence combination?
   A. 3 Tesla, gradient recalled echo
   B. 3 Tesla, spin-echo echo-planar
   C. 1.5 Tesla, gradient recalled echo
   D. 1.5 Tesla, spin-echo echo-planar

Correct Answer: D

Rationale:
The presence of liver iron can diminish MR signal and prevent successful tracking of hepatic shear waves. In this setting, there is greater signal loss at 3 Tesla vs. 1.5 Tesla, and when using a gradient recalled echo sequence vs. a spin-echo echo-planar sequence. Thus, MR elastography in patients with excess liver iron is ideally performed at a field strength of 1.5 Tesla using spin-echo echo-planar imaging.

Reference

MRI of Common Pediatric Liver Lesions
Prakash Masand, MD

7. Malignant transformation of Hepatocellular adenoma to Hepatocellular carcinoma is rarely reported, but is an accepted risk, particularly when the diameter of the adenoma exceeds...?
   A. 1 cm
   B. 3 cm
   C. 5 cm
Correct Answer: C

Rationale:
Malignant transformation of HCA to HCC is rarely reported, but is an accepted risk, particularly when the diameter of the adenoma exceeds 5 cm [Stoot et al. 2010; Grazioli et al. 2013]. In a systematic review, Stoot and colleagues reported an overall frequency of malignant transformation of 4.2% for HCAs [Stoot et al. 2010] (67 of 1635 HCAs, CI 0–100%).

Only three cases showed malignant transformation for tumors <5 cm in diameter, which represented 4.4% of the total number of HCCs arising from HCAs (3 out of 67).

Reference:

8. The histochemical surface marker GLUT 1 (glucose transporter protein) is expressed by which benign vascular tumor...
A. Rapidly involuting congenital hepatic hemangioma
B. Infantile hepatic hemangioma
C. Tufted angioma
D. Non involuting congenital hepatic hemangioma

Correct Answer: B

Rationale:
Infantile hepatic hemangiomas are vascular tumors that involve the proliferation of benign endothelial-like cells that possess histochemical markers (GLUT-1, Lewis Y antigen, FcyRII, and merosin); these markers are also present on placental blood vessels. The immunohistochemical profile differentiates IH from other vascular birthmarks or tumors.

The pathophysiology associated with the unique natural history of these lesions, with initial rapid proliferation followed by gradual involution and regression, has not been completely elucidated. One etiologic hypothesis speculates that cells are “embolized” from the placenta. Another suggests that infantile hemangiomas result from somatic mutations in a gene mediating endothelial cell proliferation. Recent data suggest an endothelial progenitor cell as the source of origin of the tumors. It has been speculated that hypoxia, either systemically (eg, due to placental insufficiency) or in a specific “niche” area of poorly perfused tissue stimulates endothelial progenitor cells to proliferate inappropriately.

Reference:
Hepatocyte Specific Contrast Material – Adenoma vs. Focal Nodular Hyperplasia vs. Other – Where Do We Stand?
Andrew Trout, MD

9. Which of the following lesions is most likely to be iso- to hyperintense on the hepatobiliary phase when imaged with a hepatocyte specific contrast material?
   A. Hepatocellular carcinoma
   B. Inflammatory adenoma
   C. Metastasis
   D. HNF-1α inactivated adenoma

Correct Answer: B

Reference:

10. Hepatobiliary phase hypointensity of a lesion imaged with a hepatocyte specific contrast material can be explained by which of the following processes:
   A. Decreased OATP expression
   B. Abnormal background liver
   C. Rapid drainage of bile from the lesion
   D. Increased blood flow to the lesion

Correct Answer: A

Reference: