

# Cirrhosis and Portal Hypertension in the Pediatric Population

Catherine A. Chapin, MD, Lee M. Bass, MD\*

## KEYWORDS

- Cirrhosis • Portal hypertension • Esophageal varices • Ascites • Biliary atresia
- Children

## KEY POINTS

- Cirrhosis is a complex diffuse process whereby the architecture of the liver has been replaced by structurally abnormal nodules due to fibrosis.
- Portal hypertension is characterized by a hepatic venous pressure gradient (HVPG) greater than 5 mm Hg with complications, such as ascites and varices, occurring at an HVPG greater than 10 mm Hg.
- Common causes of portal hypertension in children include extrahepatic portal vein obstruction, biliary atresia, alpha 1 antitrypsin deficiency, and autoimmune hepatitis, among others.
- Gastrointestinal bleeding secondary to esophageal or gastric varices may present as hematemesis or melena. Vasoactive drug therapy should be initiated as soon as possible before endoscopic treatment.
- Surgical shunt procedures or transjugular intrahepatic portosystemic shunt may be useful therapeutic options in patients with refractory portal hypertension.

## INTRODUCTION

Cirrhosis is defined by the World Health Organization as a diffuse process whereby the architecture of the liver has been replaced by structurally abnormal nodules due to fibrosis.<sup>1</sup> Cirrhosis is a common outcome of a wide spectrum of disease processes (Table 1). The pathophysiology of cirrhosis is complex and involves a dynamic interplay between hepatocyte injury, cellular response to injury, and regeneration.<sup>2</sup> Fibrosis

---

Conflict of Interest Statement: Neither Dr L.M. Bass nor Dr C.A. Chapin have any financial conflicts pertaining to the subject matter of this article.

Division of Pediatric Gastroenterology, Hepatology and Nutrition, Ann & Robert H. Lurie Children's Hospital of Chicago, Northwestern University Feinberg School of Medicine, 225 East Chicago Avenue, Box #65, Chicago, IL 60611, USA

\* Corresponding author.

E-mail address: [lbass@luriechildrens.org](mailto:lbass@luriechildrens.org)

Clin Liver Dis ■ (2018) ■-■

<https://doi.org/10.1016/j.cld.2018.06.007>

1089-3261/18/© 2018 Elsevier Inc. All rights reserved.

[liver.theclinics.com](http://liver.theclinics.com)

**Table 1**  
**Causes of cirrhosis and/or portal hypertension in children**

<b>Type</b>	<b>Disorders</b>
Genetic-metabolic disorders	$\alpha$ 1-Antitrypsin deficiency <b>Amyloidosis</b> Bile acid synthesis defects Cystic fibrosis Galactosemia Gaucher disease Glycogen storage disease type III and IV Hepatic porphyrias Hereditary fructose intolerance Hereditary hemochromatosis Indian childhood cirrhosis Langerhans cell histiocytosis Mitochondrial hepatopathies Niemann-Pick disease type C <b>Sarcoidosis</b> Tyrosinemia type I Wilson disease Wolman disease (lysosomal acid lipase deficiency)
Infectious diseases	Ascending cholangitis Chronic hepatitis B $\pm$ delta virus Chronic hepatitis C Cytomegalovirus Hepatitis E Herpes simplex virus Recurrent neonatal sepsis Rubella <b>Schistosomiasis</b> <b>Tuberculosis</b>
Inflammatory diseases	Autoimmune hepatitis Primary sclerosing cholangitis
Cholestatic diseases and biliary malformations	Alagille syndrome and nonsyndromic bile duct paucity Bile duct stenosis Biliary atresia Choledochal cyst Congenital hepatic fibrosis Caroli disease (intrahepatic cystic biliary dilatation) Progressive familial intrahepatic cholestasis
Vascular lesions	<b>Arteriovenous fistula</b> Budd-Chiari syndrome Congenital cardiomyopathy <b>Congenital stenosis or extrinsic compression of the portal vein</b> Congestive heart failure Constrictive pericarditis <b>Nodular regenerative hyperplasia</b> <b>Portal vein thrombosis</b> Sinusoidal obstructive syndrome <b>Splenic vein thrombosis</b> Venocaval web/inferior vena cava obstruction
Drugs and toxins	Hepatotoxic drugs (isoniazid, methotrexate) Hypervitaminosis A Natural toxins (eg, mushrooms) Organic solvents <b>Peliosis hepatis</b> (anabolic steroids, azathioprine) Total parenteral nutrition

(continued on next page)

Download English Version:

<https://daneshyari.com/en/article/11022081>

Download Persian Version:

<https://daneshyari.com/article/11022081>

[Daneshyari.com](https://daneshyari.com)