



Entrustable Professional Activities

EPA 2 for Pediatric Gastroenterology

EPA 2: Care of Infants, Children, and Adolescents with Acute and Chronic Liver Diseases, Biliary/Cholestatic Diseases, Pancreatic Disorders, and Those Requiring Liver Transplantation

Supervision Scale for This EPA

1. Trusted to observe only
2. Trusted to execute with direct supervision and coaching
3. Trusted to execute with indirect supervision and discussion of information conveyed for most simple and some complex cases
4. Trusted to execute with indirect supervision but may require discussion of information conveyed for a few complex cases
5. Trusted to execute without supervision

Description of the Activity

Practicing gastroenterologists/hepatologists must be trained to care for children and adolescents with acute and chronic hepatobiliary disease as well as liver transplantation and pancreatic disorders. Pediatric gastroenterologists/hepatologists need to be familiar with current knowledge of disease processes and presentations. Additionally, subspecialists should be able to manage acute issues as well as long-term chronic management including the transition of care.

The specific functions which define this EPA include:

1. Understanding and applying the epidemiology, pathophysiology, pathogenesis, and natural history of acute/chronic liver diseases, biliary diseases, liver transplantation, and pancreatic disorders
2. Interpreting and applying basic/translational and clinical research in hepatology, biliary, and pancreatic disorders to the care of patients
3. Recognizing and diagnosing children with suspected liver, biliary, and pancreatic disorders in a variety of clinical presentations
4. Managing children and adolescents with liver, biliary, pancreatic disorders, and liver transplantation including the acute presentations and emergencies as well as long-term management of complex chronic diseases
5. Educating parents and children on liver, biliary, and pancreatic diseases including cause, treatment, and clinical course
6. Leading, directing and coordinating care for children/adolescents with liver, biliary, and pancreatic diseases within the medical system and the community

Judicious Mapping to Domains of Competence

- Patient Care
- Medical Knowledge
- Practice-Based Learning and Improvement
- Interpersonal and Communication Skills



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- Professionalism
- Systems-Based Practice
- Personal and Professional Development

Competencies Within Each Domain Critical to Entrustment Decisions

PC 5:	Performing complete physical exams
PC 9:	Counseling patients and families
MK 1:	Demonstrate knowledge
MK 2:	Practicing EBM
PBLI 2:	Setting goals
PBLI 8:	Developing teaching skills
PBLI 10:	Engaging in lifelong learning
ICS 2:	Demonstrating insight into emotion
PPD 8:	Dealing with uncertainty

Context for the EPA

Rationale/Scope of Practice: Pediatric gastroenterologists care for patients with acute and chronic liver and biliary disorders, some of which may require liver transplantation, and pancreatic disorders.

Acute and Chronic Hepatobiliary Diseases: Practicing subspecialists must be trained to care for children and adolescents with acute and chronic liver disease. Within the pediatric gastroenterology subspecialty, these relatively uncommon conditions account for a significant amount of morbidity and mortality. Pediatric gastroenterologists need to be familiar with current knowledge of liver disease processes. Additionally, subspecialists should be able to manage acute issues as well as long-term management of chronic illnesses including the transition of care. The diagnosis and treatment of acute and chronic liver disease requires the knowledge and understanding of anatomy, pathophysiology, epidemiology, and diagnosis/management of these diseases that include acute liver failure, chronic liver failure, viral hepatitis, autoimmune hepatitis, fatty liver disease, congenital liver diseases, hepatic tumors, infectious/drug-induced hepatitis, other liver conditions.

The diagnosis and treatment of diseases of the biliary tree, cholestatic liver disease, and metabolic liver disease compose a significant proportion of practice for pediatric gastroenterologists. Due to the relative rarity of these conditions, pediatric gastroenterologists serve a key role as consultants to general pediatricians who encounter patients with signs/symptoms (e.g., jaundice, right upper quadrant (RUQ) pain) or findings (laboratory or radiographic abnormalities) suggestive of biliary/liver pathology. Pediatric gastroenterologists should be able to obtain key diagnostic information from patient history, physical examination and targeted studies (including labs, imaging, and procedures) to evaluate for biliary and liver pathology. Pediatric gastroenterologists planning clinical practice may choose to obtain proficiency with percutaneous liver biopsy to diagnose hepatobiliary pathology although not required, and some pediatric gastroenterologists may also work towards proficiency in endoscopic retrograde cholangiopancreatography (ERCP), though for most, the key proficiency will be appropriate referral to a center that performs ERCP on children. Additionally, pediatric gastroenterologists must provide longitudinal care for patients with disorders of diseases of the bile ducts, cholestatic liver disease, and metabolic liver disease. The pediatric gastroenterologist may serve as the point person in management in some of these rare diseases (e.g., biliary atresia, progressive familial intrahepatic cholestasis (PFIC), Alagille syndrome, biliary atresia, Crigler-Najjar



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syndrome, glycogen storage disease, and others), whereas they will be a part of a multidisciplinary team approach for others (e.g., cystic fibrosis, alpha-1 antitrypsin deficiency, mitochondrial disease).

Liver Transplantation: Pediatric gastroenterologists participate in the care of children and adolescents who will have or have undergone a liver transplantation. Pediatric gastroenterologists should have a complete understanding of acute and chronic liver conditions requiring transplantation. Pediatric gastroenterologists should be familiar with the indications for liver transplantation and participate in the pre-transplant workup and preparation as well as post-transplant care.

Pancreatic Diseases: Pediatric gastroenterologists must understand acute and chronic pancreatic diseases, including hereditary disorders that occur in the pediatric population. Pediatric gastroenterologists must be able to diagnose and treat pancreatic disorders and their complications, as well as demonstrate a thorough understanding of their pathophysiology. Pediatric gastroenterologists should have a thorough understanding of the spectrum of pancreatic disease including epidemiology, etiology, pathophysiology, natural history, and disease management. These diseases include acute, acute recurrent, and chronic pancreatitis, including hereditary pancreatitis, autoimmune pancreatitis, pancreatitis due to toxic/metabolic causes, and pancreatitis due to anatomical variants. Genetic and metabolic pancreatic disorders that can affect both the endocrine and exocrine pancreatic function including cystic fibrosis, Schwachman-Diamond, and Johanson-Blizzard as well as other disorders of pancreatic dysfunction are also important considerations. In addition, pediatric gastroenterologists must be familiar with the normal anatomy and embryology of the pancreas including structural variants (e.g., pancreatic divisum, annular pancreas, and pancreatic agenesis).

Pediatric gastroenterologists must be able to care for pediatric patients of all ages with these conditions and unite a team of caregivers to provide exceptional patient care and must display skills of communication and lifelong learning skills. Pediatric gastroenterologists must be sensitive to a wide variety of patient's backgrounds and beliefs in their caring for children with gastrointestinal disorders and must demonstrate a caring and empathetic attitude towards patients and families.

Curricular Components That Support the Functions of the EPA

1. Understanding and applying the epidemiology, pathogenesis, and natural history of acute/chronic liver diseases, biliary diseases, and pancreatic disorders
 - Acute/Chronic Hepatobiliary Diseases
 - Knows and understands the basic science of the acute and chronic liver disease etiologies including pathogenesis, genetics, and immunology
 - Understands the natural history of acute and chronic liver diseases
 - Understands the normal physiology of the liver, and understands liver enzyme and liver function tests and their abnormalities in disease
 - Demonstrates understanding of liver metabolic pathways, such as the urea cycle, glycogen synthesis, and breakdown
 - Demonstrates knowledge of basic biliary anatomy and bile duct development
 - Differentiates between gallbladder/biliary infection and inflammation and directs management accordingly
 - Describes the normal physiology of bilirubin and bile acid metabolism, including synthesis, conjugation, excretion and recirculation, and factors that influence bile flow



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- Liver transplantation
 - Knows and understands the clinical science of liver transplantation including indications, epidemiology, natural history, nutritional, gastrointestinal and surgical management, and complications
 - Knows and understands the basic science of liver transplantation including immunology, genetics, post-transplant physiology, and common disease processes after transplantation
 - Knows and understands the mechanism of action, side effects, and other complications of medications and treatments used before and after liver transplantation including steroids, immunomodulators, biologics, antivirals, and other medications
 - Knows and understands the acute and chronic immunological consequences of liver transplantation including acute and chronic rejection, autoimmune post-transplant processes, and other processes in relation to graft vs. host reactions
 - Pancreatic disorders
 - Knows and understands the normal anatomy of the pancreas and congenital variants
 - Knows and understands the physiology of pancreatic exocrine secretion of digestive enzymes, including the types of enzymes, their mechanisms of activation, regulation, and roles in digestion
 - Demonstrates knowledge of established and evolving biomedical, clinical, and epidemiological sciences of pancreatic disorders, as well as the application of this knowledge to patient care
 - Knows and understands the epidemiology, etiology, pathophysiology, natural history, and management of acute and chronic pancreatitis
 - Demonstrates a basic understanding of the molecular genetics of pancreatic disease including but not limited to gallstone pancreatitis, hereditary pancreatitis and cystic fibrosis, their diagnosis, and management
2. Interpreting and applying basic/translational and clinical research in hepatology, biliary, and pancreatic disorders to the care of patients
- Acute/chronic hepatobiliary diseases
 - Applies evidence-based medicine to the care of children with liver disease
 - Participates in lifelong learning as it relates to care of children with liver disease
 - Demonstrates use of available evidence to evaluate and optimize the care of patients with acute and chronic liver disease, biliary disorders, cholestatic liver disease, and/or metabolic liver diseases
 - Knows and understands principles of evidence-based medicine as applied to the management of hepatobiliary and metabolic disorders
 - Liver transplantation
 - Knows and understands research as it applies to liver transplantation, management pre- and post-transplantation, and transplant rejection
 - Applies evidence-based medicine to the care of children with liver transplantation
 - Knows and understands medications used in transplantation



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- Pancreatic disorders
 - Demonstrates use of available evidence to investigate, evaluate, and improve the care of patients with pancreatic disorders
 - Knows and understands principles of evidence-based medicine as applied to children with pancreatic disorders
 - Knows and understands that clinical practice guidelines are suggestions for clinical care and may be flexible and evolve with time
 - Explains clinical decisions in the context of evidence-based medicine
 - Demonstrates knowledge of research that has been performed in patient care, diagnosis, and pathophysiology in children with pancreatic disorders

- 3. Recognizing and diagnosing children with suspected liver, biliary, and pancreatic disorders in a variety of clinical presentations
 - Acute/chronic hepatobiliary diseases
 - Articulates the differential diagnosis of neonatal cholestasis and formulates an appropriate evaluation and treatment plan
 - Demonstrates proficiency in the evaluation and treatment of infants, children, and adolescents with cholestasis, including indications for genetic testing and available treatment options for pruritus. Knows the indications for surgical intervention, such as biliary diversion or liver transplantation in this patient population
 - Knows and understands the initial diagnostic evaluation for metabolic liver disease and coordinates care of these patients in consultation with the other medical professionals
 - Gathers essential and accurate information regarding risk factors such as intravenous injections, foreign travel, contact with jaundiced individuals, medications/toxin exposures, and sexual history in adolescents if sexually active
 - Gathers essential and accurate information from the patient/parent, focused on concerning signs and symptoms for potential hepatobiliary pathology (jaundice, acholic stool, failure to thrive, RUQ pain, pruritus, encephalopathy, etc.)
 - Performs an initial appropriate physical examination with evaluation for evidence of chronic liver disease including hepatomegaly, splenomegaly, ascites, and a complete neurological exam for evaluation of encephalopathy
 - Performs an appropriate initial diagnostic evaluation for a child with acute and chronic liver disease (including laboratory, imaging, and histological interpretation). Understands that the etiology for acute liver failure (ALF) is usually age dependent and thus the need for tailoring of initial workup
 - Performs an appropriate initial diagnostic for a child with suspected biliary disease or cholestatic liver disease (laboratory testing and imaging)
 - Explains the differences between acute and chronic liver disease
 - Demonstrates knowledge of the differential diagnosis for acute (e.g., acute infectious hepatitis, drug/toxin hepatitis, [ALF]) and chronic liver disease (e.g., chronic viral hepatitis, autoimmune hepatitis, nonalcoholic steatohepatitis, congenital liver disease), including clinical presentation and features, epidemiology, natural history, and treatment (including liver transplantation)
 - Recognizes the clinical and laboratory findings of a child in ALF and explains the management of ALF (including PICU support, prevention/treatment of complications, hepatic support, and consideration for liver transplantation)



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- Liver transplantation
 - Performs an appropriate initial workup for a child with liver or metabolic disease who may be a suitable candidate for liver transplantation, including laboratory tests, appropriate endoscopic evaluation, liver biopsy and pathology review, imaging, psychological and socio-economic evaluations
 - Appropriately contacts a hepatology transplant center for referral when indicated if located in a nontransplant center
 - Obtains a comprehensive history and physical exam focused on the appropriate systems for post-transplant children and adolescents
- Pancreatic disorders
 - Obtains a detailed, complete, and accurate history of pancreatic disorders and presentation of common pancreatic disorders such as acute and chronic pancreatitis as well as disorders of pancreatic exocrine dysfunction
 - Performs a physical exam that would appropriately identify signs of pancreatitis and/or pancreatic insufficiency and related systemic manifestations
 - Accurately assesses nature, acuity, and severity of the clinical problem
 - Provides a comprehensive assessment of the medical issues that must be addressed for each patient
 - Orders necessary and appropriate laboratory and diagnostic tests in light of the clinical presentation and formulates a treatment plan based on the diagnosis
- 4. Managing children and adolescents with liver, biliary, and pancreatic disorders including the acute presentations and emergencies as well as long-term management of complex chronic diseases
 - Acute/chronic hepatobiliary diseases
 - Establishes the diagnosis, stages the disease, and detects complications of liver disease
 - Develops an initial treatment plan with monitoring of medications and potential side effects
 - Recognizes remission and exacerbation in chronic liver disease and develops a treatment plan for disease exacerbations and maintenance therapy
 - Identifies patients in need of interventional diagnostics for hepatobiliary disease (including liver biopsy, endoscopic retrograde cholangiopancreatography [ERCP], percutaneous transhepatic cholangiogram) and describe the risks and benefits of each procedure
 - Recognizes clinical signs and symptoms in a patient presenting with acute or chronic liver disease that may be indicative of an underlying metabolic disorder
 - Monitors for and recognizes complications of both acute and chronic liver disease
 - Performs endoscopy for esophageal varices if indicated (see Gastroenterology EPA 5)
 - Distinguishes among the various imaging modalities for liver disease including but not limited to abdominal radiography, ultrasonography with doppler, CT scans and MRI, including magnetic resonance cholangiopancreatography (MRCP) and/or ERCP and chooses the appropriate modality based on top diagnostic considerations
 - In consultation with pathology, correctly interprets liver biopsy findings, including identification of findings suggestive of biliary obstruction or metabolic liver disease
 - Knows and understands the use of treatment modalities for liver disease including
 - Steroids



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- Immunomodulators
 - Biologics
 - Anti-virals
 - Oral, enteral and parenteral nutrition
 - Other medications (vitamins E, D, K, fish oil, etc.)
 - Pharmacotherapy in portal hypertension (e.g., Beta-blockers, octreotide)
 - Endoscopic or surgical treatment for portal hypertension
 - Aldosterone antagonist and diuretics for ascites
 - Albumin infusion
 - Pharmacotherapy for hepatic encephalopathy (e.g., lactulose, antibiotics)
 - Nonconventional therapies, alternative medicine
 - Liver transplantation
 - Evolving therapies / new treatment modalities for hepatic support (e.g., Molecular Absorbent Recirculating Systems, hepatocyte transplantation)
- Recognizes extra-intestinal manifestations of liver disease
 - Interprets pathology findings
 - Knows and understands health care maintenance and vaccinations in children with chronic liver disease and immunosuppression medical regimens
 - Recognizes and addresses psychosocial concerns
 - Explains the mechanism of action, side effects, and other complications of medications and treatments for chronic liver disease including immunosuppression medical regimens, nutritional and vitamin supplementation, and other medications
 - Knows and understands the extrahepatic manifestation of liver disease, including but not limited to neurological involvement, renal disease, and hematological disorders
 - Demonstrates knowledge of and recognizes complications of liver disease, including malnutrition and growth failure, pruritus, ascending cholangitis, portal hypertension, ascites, variceal bleeding, coagulopathy, hepatic encephalopathy, hepatorenal syndrome, hepatopulmonary syndrome, bacterial infections (spontaneous bacterial peritonitis), and hepatocellular carcinoma
 - Recognizes signs and symptoms of End Stage Liver Disease (ESLD) and appropriate timing to refer for liver transplantation
 - Identifies liver metabolic diseases that may lead to evaluation for liver transplantation
 - Demonstrates knowledge regarding transmission and prevention strategies for the viral hepatitis
 - Applies quality measures to improve care of children with liver disease
- Liver transplantation
 - Accurately assesses nature, acuity, and severity of the clinical post-transplant problems
 - Utilizes laboratory test, imaging studies, and pathologic data to monitor patient post-transplant
 - Knows and understands the short- and long-term consequences of immunosuppressive therapy including steroids, immunomodulators, biologic/antibody therapy, and other emerging therapies
 - Recognizes acute and chronic post-transplant complications: nutritional, metabolic, infectious, and surgical
 - Utilizes clinical practice guidelines in the care of post-transplant children



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- Pancreatic disorders
 - Performs continuing evaluation of patients to evaluate safety and efficacy of observations and treatment plan as instituted; orders additional studies and/or adjusts therapy as clinically indicated
 - Develops and carries out patient management plans and communicates with patient, family, primary as well as consulting physician, resident physicians, nursing staff, nutrition support team, and other ancillary health service providers
 - Knows and understands the indications for and interprets laboratory results in the diagnosis and management of pancreatic diseases
 - Knows and understands the principles, utility, indications for, and basic interpretation of all radiographic studies of the pancreas including their potential risks and benefits
 - Recognizes therapeutic options for the treatment of pancreatic disease, including the role of nutritional therapy, pain management, and endoscopic, radiographic, and surgical intervention
 - Recognizes the indications for invasive testing of the pancreas including endoscopic ultrasound (EUS), ERCP, and endoscopic management of pancreatic diseases

- 5. Educating parents and children on liver, biliary, and pancreatic diseases including cause, treatment, and clinical course.
 - Acute/chronic hepatobiliary diseases
 - Educates parents and children on cause, treatment, and other aspects of liver disease, including maintenance health check and cancer surveillance
 - Effectively communicates disease information, treatment plan, and outcome to parents and children
 - Assesses patient's and parents' level of medical understanding and explains medical decision making in understandable terms
 - Creates a comfortable environment where parents can share sensitive history that may have influenced development of disease (toxic ingestions, consanguinity, etc.)
 - Enables patients and parents to be comfortable asking questions and making suggestions

 - Liver transplantation
 - Empathetically discusses the process of evaluation for liver transplantation and the ethics of the national organ donation system
 - Involves patient and their families in creating treatment plans
 - Ensures that the patient and their family understand the rationale for the plan of care
 - Encourages patient to freely discuss their care and concerns about medications and current management by exhibiting empathetic listening skills
 - Ensures that patients understand the correct way to administer their medications

 - Pancreatic disorders
 - Provides education to patients and families concerning the pathophysiology and manifestations of their gastrointestinal disease process. Discusses the proper use of prescribed medications and/or interventions (e.g., ERCP) including potential adverse effects
 - Effectively communicates disease information, treatment plan, and outcome to patients and their families



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6. Leading, directing, and coordinating care for children/adolescents with liver, biliary, and pancreatic diseases within the medical system and the community
 - Chronic GI disease (in general)
 - Recognizes the importance of a biopsychosocial model of care for patients with chronic disease that addresses not only the physical but social, emotional, behavioral, and mental health issues that often complicate chronic illness
 - Ensures that the patient and family are part of the interdisciplinary health care team and engages them in shared decision making
 - Acknowledges the anxiety that accompanies diagnosis and management of chronic disease and supports the patient/ family's emotional response to living with uncertainties through ongoing updates as knowledge becomes available and balancing difficult conversations with hope
 - Recognizes issues that may contribute to poor compliance or noncompliance and is able to recruit necessary resources to address these issues (i.e., health educator for health literacy issues, psychiatry/social work for depression)
 - Acute/chronic hepatobiliary diseases
 - Coordinates care of children with acute and chronic hepatobiliary disease including PCPs, surgeons, psychologists/social workers, nutritionists, radiologists, and other medical professionals
 - Effectively communicates with other medical professionals involved in the care of patients with acute and chronic liver disease
 - Prepares for transition and appropriately transitions care in children with liver disease, biliary disease, and metabolic disorders to adult gastroenterologists/hepatologists
 - Communicates effectively with all members of a multidisciplinary team, including nursing staff, medical assistants, pharmacy staff, social workers, primary care providers, and other consultants (surgeons, radiologists, pathologists)
 - Practices high-quality, fiscally responsible and cost-effective health care by demonstrating consideration of costs to the patients' families and the system in recommending diagnostic tests, treatments, and follow-up management
 - Advocates for patients within the health care system, particularly those in resource-poor settings
 - Knows and understands the link between early and effective management of congenital hepatobiliary disease (e.g., biliary atresia, cystic fibrosis, primary hyperoxaluria) and prevention of secondary health care costs later in life
 - Educates health care professionals and the lay public on issues in management of hepatobiliary disease and acts as an advocate for recognition of disease (red flags) and management (best practices)
 - Liver transplantation
 - Communicates management plans, in a timely fashion, to the patient, family, primary physician, resident physicians, nursing staff, nutrition team, and other ancillary health service providers
 - Demonstrates an awareness of and responsiveness to the larger context and system of health care and the ability to effectively call on system resources to provide care that is of optimal value
 - Practices high-quality fiscally responsible and cost-effective health care by demonstrating consideration of costs to the patients' families and the system in recommending diagnostic tests, treatments, and follow-up management



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- Advocates for patients within the health care system
- Coordinates care of post-transplant children with primary care physicians, surgeons, psychologists, social workers, dietitians, radiologists, and other medical professionals
- Transitions care of post-transplant children to adult gastroenterologists
- Pancreatic Disorders
 - Effectively communicates with other medical professionals involved in the care of the patient
 - Works effectively as a member or leader of a health care team coordinating care of the patient

Problems that **generally** require further consultation where the role of the subspecialist is to recognize and provide preliminary evaluation and refer/co-manage

- Liver Transplantation
- Endoscopic retrograde cholangiopancreatography (ERCP)
- Cholecystectomy
- Surgery for hepatic masses or other lesions
- Psychiatry/Psychology/Pain Medicine/Social work
 - Anxiety/Depression
 - Chronic pain

EPA Authors

Cary G. Sauer, MD, M.Sc. (lead), Keith Benkov, MD, Carolina S. Cerezo, MD, FAAP, Jeannie S. Huang, MD MPH, Alan M. Leichtner, MD MSHPEd, Kathleen M. Loomes, MD, Michael R Narkewicz, MD, Rubén E. Quirós-Tejeira, MD, Jacob Robson, MD, Yumirle Turmelle, MD, Toba Weinstein, MD

Curricular Components Authors

Cary Sauer, MD, Jacob Robson, MD, Caroline Cerezo, MD, Yumirle Turmelle, MD, Kathleen Loomes, MD, Jeannie Huang, MD, Keith Benkov, MD, Ruben Quiros, MD, Jonathan Tietelbaum, MD, Toba Weinstein, MD, Alan Leichtner, MD, Michael Narkewicz, MD