

Entrustable Professional Activity

1. Title: Care for infants, children, and adolescents with disorders of biliary tract disease, cholestatic liver disease, and metabolic liver disease
2. Description of Activity:

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 symptoms (eg jaundice, right upper quadrant pain) or findings (abnormal Alk Phos, AST/ALT, GGT, Bilirubin, abnormal findings on ultrasound) suggestive of biliary/liver pathology. With the increased trend towards subspecialty referral for issues other than well child care, gastroenterologists are often asked to see these patients in both the outpatient and inpatient settings, getting involved early to help guide diagnostic workup and initial therapeutic interventions. At the completion of training, a pediatric gastroenterologist should be able to obtain key diagnostic information from patient history, physical examination and targeted studies (including labs, imaging, procedures) to evaluate for biliary and liver pathology. Pediatric gastroenterologists planning clinical practice should obtain proficiency with percutaneous liver biopsy to diagnose hepatobiliary pathology, and some pediatric gastroenterologists may also work towards proficiency in 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 (biliary atresia, PFIC, Alagille syndrome, biliary atresia, Criglar-najjar syndrome, glycogen storage disease, and others), whereas they will be a part of a multidisciplinary team approach for others (cystic fibrosis, alpha-1 antitrypsin deficiency, mitochondrial disease).

The functions of this activity include:

1. Understand the epidemiology, pathogenesis and natural history of biliary tract disease, cholestatic liver disease and metabolic liver disease.
2. Understand the indications for ERCP and/or cholecystectomy in patients presenting with biliary tract disease.
3. Understand the differential diagnosis, evaluation and management of neonatal cholestasis. This includes proficiency in interpretation of liver biopsy and indications for surgical consultation and intra-operative cholangiogram.
4. Understand the diagnostic evaluation for children of all ages presenting with cholestasis, including indications for genetic testing.
5. Understand the indications for testing to establish a diagnosis of metabolic liver disease, and manage these patients in collaboration with the metabolism service.
6. Understand the systemic manifestations that may be present in patients with cholestatic and metabolic liver diseases (e.g. cardiac or renal involvement in Alagille syndrome, pulmonary disease in alpha-1 antitrypsin deficiency, neurologic involvement in mitochondrial disease, etc.)
7. Understand appropriate indications and contraindications for liver transplantation in these patient populations.
8. Manage the care of children and adolescents with biliary tract disease, cholestatic liver disease and metabolic liver disease. This includes nutritional management, repletion of fat-soluble vitamins and treatment of pruritus.
9. Educate parents and children on the management and natural history of these biliary tract and liver diseases.
10. Lead and direct care for children and adolescents with biliary tract disease, cholestatic liver disease and metabolic liver disease, including coordination of care with multiple teams of specialists.
11. Understand basic, translational and clinical research in these disorders.

3. Domains of Competence (Judicious Mapping)

- Patient Care
- Medical Knowledge and Diagnostic Skills Required (from Training Guidelines)
- Practice Based Learning
- Interpersonal Communication
- Professionalism
- Systems-based Practice
- Personal and Professional Development

4. Competencies within each domain critical to entrustment decision (From Pediatric Milestones Document)

- PC 5, 9
- MK 1, 2
- PBLI 2, 8, 10
- ICS 2
- PPD 5, 8

5. Curriculum

Knowledge:

The fellow must have a basic, broad understanding of all that goes into a comprehensive assessment of a patient with hepatobiliary pathology. This starts with an understanding of the anatomy of the biliary tree, the physiology of biliary system and key metabolic pathways housed in the liver. From this, trainees will develop the ability to recognize congenital and acquired structural biliary abnormalities, abnormalities in bilirubin metabolism (production, conjugation, recirculation) and movement (hormonal regulation, gallbladder motility, cholestasis/obstruction) and malfunction of key liver synthesis/storage and breakdown pathways. With this basic knowledge of anatomy and physiology, the trainee should demonstrate proficiency at identifying pathology related to: 1) anatomic biliary/gallbladder disease (such as biliary atresia, choledochal cyst, cholelithiasis), 2) infection/inflammation of the biliary tree/gallbladder (such as ascending cholangitis, primary and secondary sclerosing cholangitis), 3) congenital issues with bilirubin metabolism and excretion (such as Crigler-Najjar, Dubin-Johnson and Gilbert's syndrome), 4) inherited cholestatic disorders (such as Alagille syndrome, PFIC, cystic fibrosis, bile acid synthetic defects) and 5) metabolic liver disorders (such as Wilson disease, alpha-1 antitrypsin deficiency, mitochondrial disorders, urea cycle defects, glycogen storage diseases, etc). An understanding of the anatomy and pathophysiology of these disorders informs creation of diagnostic and therapeutic management plans. At the end of training, the fellow must be proficient in current best practices in evaluation (including judicious use and interpretation of lab tests, imaging modalities and diagnostic procedures) and management (watchful waiting, medications, procedural intervention, liver transplantation) for the above categories of conditions.

Skills:

The fellow must be able to efficiently obtain a focused medical history from patient and/or parents, using appropriate questions and lay terminology to help patients and caregivers accurately summarize pertinent points (jaundice, pain, stool color, etc). This history must then be combined with objective data from the physical examination and diagnostic workup (laboratory values, imaging studies, procedures) to refine a differential diagnosis for the suspected hepatobiliary pathology. Specifically, the fellow should have an understanding of the differential utility of ultrasound, CT and MRI for the evaluation of hepatobiliary pathology and be able to decide when to move on to liver biopsy or invasive

imaging/intervention via ERCP. With these data, the fellow should be able to outline a clear, feasible plan for intervention (which may include reassurance, close monitoring, further tests/imaging/procedures, medication, procedural intervention, or even liver transplant evaluation), allowing the family to ask questions and express their concerns about potential complications or side effects of the management plan. Clear documentation of the conversation must be provided to the patient and caretakers and plans for follow-up should always be set before the end of any visit.

Attitudes:

In speaking with families about hepatobiliary issues, the fellow should recall that while knowledge on these topics is germane to their field, it is often a topic that is not commonly understood by the lay public, and may require extensive explanation and multiple re-visitations over time. The fellow must understand the ethics of liver transplant/organ donation and display sensitivity in discussing many of the congenital/inherited disorders of bilirubin/liver metabolism. The fellow should be comfortable working on a multidisciplinary team in generating a differential diagnosis and devising a treatment plan—including radiologists, pathologists, surgeons and others (oncologists, genetics/metabolism, etc). To allow for entrustment to unsupervised practice, the trainee must show the willingness to patiently accept information from patients, synthesize with additional data, and then respond with a comprehensive management plan which takes into account and balances many key factors, including underlying disease process, patients/caretaker understanding of pathology, socioeconomic status, cultural beliefs, end of life decisions, etc.

Competency-based objectives

Patient Care

1. Gather essential and accurate information from the patient/parent, focused on red flags for potential hepatobiliary pathology (jaundice, acholic stool, failure to thrive, RUQ pain, pruritus, encephalopathy, etc.)
2. Perform a directed physical examination for a patient with suspected hepatobiliary disease and interpret results accordingly based on pre-test probability of findings
3. Perform an appropriate initial diagnostic for a child with suspected hepatobiliary disease (laboratory testing and imaging)
4. Identify patients in need of interventional diagnostics for hepatobiliary disease (liver biopsy, ERCP, percutaneous transhepatic cholangiogram) and describe the risks and benefits of each procedure.
5. In consultation with pathology, correctly interpret liver biopsy findings, including identification of findings suggestive of biliary obstruction or metabolic liver disease.
6. Understand imaging modalities for biliary and liver metabolic pathology, including but not limited to abdominal radiography, CT scan, ultrasonography, MRI/MCRP, HIDA scan and ERCP
7. Identify biliary and liver metabolic diseases that may lead to evaluation for liver transplantation

Medical Knowledge

1. Demonstrate knowledge of basic biliary anatomy and bile duct development
2. Demonstrate understanding of liver metabolic pathways, such as the urea cycle, glycogen synthesis and breakdown.
3. Differentiate between gallbladder/biliary infection and inflammation and direct management accordingly
4. Describe the normal physiology of bilirubin and bile acid metabolism, including synthesis, conjugation, excretion and recirculation, and factors that influence bile flow.
5. Understand the differential diagnosis of neonatal cholestasis, and formulate an appropriate evaluation and treatment plan.

6. Demonstrate proficiency in the evaluation and treatment of infants, children and adolescents with cholestasis, including indications for genetic testing and available treatment options for pruritus. Understand indications for surgical intervention, such as biliary diversion, or liver transplantation in this patient population.
7. Understand the initial diagnostic evaluation for metabolic liver disease, and coordinate care of these patients in consultation with the metabolism service.

Practice Based Learning

1. Demonstrate use of available evidence to evaluate and optimize the care of patients with biliary disorders, cholestatic liver disease and/or metabolic liver diseases
2. Understand principles of evidence-based medicine, as applied to the management of hepatobiliary and metabolic disorders
3. Balance the cost of evaluation with the benefit of early diagnosis and treatment for rare hepatobiliary metabolic diseases (PFIC or Alagille syndrome) based on an understanding the epidemiologic principles of positive predictive value
4. Understand the concept of cost-benefit analysis, for both outpatient clinic management and intensive inpatient therapy
5. Understand that clinical practice guidelines are suggestions for clinical care and may be flexible and evolve with time
6. Interact with faculty and colleagues to discuss evaluations of complex patients and incorporate feedback into promoting professional growth and practice improvement

Interpersonal Skills

1. Create therapeutic relationships with patients and their care providers
2. Assess patients' and parents' level of medical understanding, recognizing for biliary and liver disease it is often minimal, and explain medical decision making in understandable terms
3. Create a comfortable environment where parents can share sensitive history that may have influenced development of disease (toxic ingestions, consanguinity, etc).
4. Enable patients and parents to be comfortable asking questions and making suggestions
5. Communicate 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)
6. Empathetically discuss the process of evaluation for liver transplantation and the ethics of the national organ donation system

Professionalism

1. Demonstrate good practices related to patient confidentiality
2. Provide emotional, social and culturally sensitive support to patients and families at various phases of the care continuum (including initial workup, refining the differential diagnosis, initiating therapy, maintenance care, and discharge from care or end of life decisions)
3. Complete all consults, medical records and patient care activities in a timely manner, and ensure good communication with team members and family
4. Respect patient's role in medical decision making and their autonomy
5. Thoughtfully discuss risks, benefits and costs of interventions with patients/parents

System-based Practice

1. Practice 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
2. Advocate for patients within the health care system, particularly those in resource poor settings
3. Understand the link between early and effective management of congenital hepatobiliary disease (e.g. biliary atresia, cystic fibrosis, primary hyperoxaluria, etc) and prevention of secondary healthcare costs later in life
4. Understand the best timing and practices for the transition of care of patients with chronic biliary or liver metabolic disease to adult providers
5. Educate health care professionals and the lay public on issues in management of hepatobiliary disease and act as an advocate for recognition of disease (red flags) and management (best practices)

Personal and Professional Development

1. Demonstrate trustworthiness that makes colleagues feel secure when the fellow is involved in key care decisions for their patients
2. Provide leadership skills that enhance the health care delivery system with the ultimate intent of improving care of patients
3. Demonstrate self-confidence that puts patients, families and members of the health care team at ease
4. Recognize that ambiguity is part of clinical medicine and respond by utilizing appropriate resources when dealing with uncertainty
5. Integrate patient's personal circumstances (cultural beliefs, economic situation, proximity to care) into construction of a health care management plan

6. Assessment Procedure

Quick Summary of EPA

End-of-Training EPA	Step 1 Description and Tasks	Step 2 Domains of Competence and Competencies within each Domain Critical to Entrustment Decisions		Step 3 Assessment Methods/Tools
<p>1. Care for infants, children, and adolescents with disorders of biliary tract disease, cholestatic liver disease, and metabolic liver disease</p>	<p>Summary:</p> <p>Pediatric gastroenterologists entering into unsupervised practice will be able to perform a comprehensive assessment of patients with suspected hepatobiliary or metabolic liver disease in a wide variety of clinical settings, and will provide diagnostic and management plans to patients and primary care providers.</p> <p>The tasks required:</p> <ul style="list-style-type: none"> • Obtain pertinent history for potential hepatobiliary pathology and perform appropriate physical exam maneuvers • Display basic knowledge of basic biliary anatomy, bilirubin metabolism and liver metabolic pathways • Direct diagnostics to refine differential diagnosis of disease, understanding costs, risks and benefits of tests and invasive procedures • Communicate management plans to patients, their families and care givers • Understand extrahepatic manifestations and indications for liver transplantation in these disorders. • Educate patients, their families and care givers, and other health professionals about diagnoses • Adapt management plan to changing clinical information • Provide consultation to referring physician and other health care providers 	Patient Care (PC)	1, 3-9	<p>Direct observations In-training examination 360 Global Ratings of Live Performance Standardized Patient Examination Portfolios</p>
		Medical Knowledge (MK)	1-2	
		Practice-Based Learning & Improvement (PBLI)	1-3, 6	
		Interpersonal & Communication Skills (ICS)	1, 3-6	
		Professionalism (P)	1-3, 5	
		Systems-Based Practice (SBP)	2, 5-6	
		Personal and Professional Development (PPD)		

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Level 1	Level 2	Level 3	Level 4	Level 5
<p>Perform history and physical exam in children presenting with red flags for hepatobiliary disease (jaundice, pruritus)</p> <p>Develop a basic initial diagnostic plan including laboratory evaluation, endoscopy, imaging and other tests as needed</p> <p>Understand biliary anatomy and liver metabolic pathways</p>	<p>Understand the natural history, epidemiology, and pathogenesis of biliary diseases</p> <p>Understand bile acid metabolism and associated disorders</p> <p>Develop differential diagnosis for biliary diseases</p> <p>Develop an appropriate initial diagnostic plan for laboratory, imaging, and procedures for biliary diseases</p> <p>Develop an appropriate diagnostic plan for neonatal cholestasis</p> <p>Recognize signs/symptoms and association abnormalities with Alagille syndrome</p> <p>Recognize BA and refer for surgical management in a timely manner</p> <p>Recognize infections and hemolytic diseases as causes of cholestatic disorders and diagnose/manage appropriately</p>	<p>Understand biliary anatomy and abnormalities present with choledochal cysts</p> <p>Understand metabolic/genetic causes of liver disease that result in cholestasis</p> <p>Recognize and manage gallbladder disease including cholelithiasis and biliary dyskinesia</p> <p>Manage patients with Alagille syndrome and understand medical/surgical options</p> <p>Educate families and children regarding biliary diseases</p> <p>Understand medication classes and side effects used to treat biliary disease</p> <p>Recognize Gilbert's syndrome and more severe disorders of bilirubin metabolism (Crigler-Najjar, Dubin-Johnson, Rotor)</p> <p>Diagnose and manage children with PFIC diseases</p>	<p>Recognize rare congenital defects of the intrahepatic bile ducts (Caroli Disease)</p> <p>Understand the molecular defects in bile acid transport and synthesis disorders (PFIC, neonatal cholestasis, bile acid synthesis defects, etc)</p> <p>Diagnose and manage biliary disorders associated with other diseases (Primary sclerosing cholangitis, CF associated biliary disease, polycystic kidney disease)</p> <p>Lead a multi-disciplinary team including dietician, surgeons, and others that care for children with biliary diseases</p> <p>Apply QI, Best Practices, and Scientific Evidence</p> <p>Transition care of patients from pediatric to adult providers</p>	<p>Participate in scholarly activity related to biliary disease</p> <p>Present research findings or clinical case at a national meeting</p> <p>Be invited to speak at regional/national meetings on biliary topics</p>

Training / Expertise Level

Entrustment Level

Execution with direct proactive supervision
 Execution with reactive supervision (on request)
 Supervision at distance post hoc supervision
 Entrustment, ready for unsupervised practice
Supervision of others junior colleagues