EPA 3: Care for Infants, Children, and Adolescents with Acute and Chronic Biliary/Cholestatic Diseases

Description of the Activity

Practicing pediatric transplant hepatologists must be trained to care for children and adolescents who have acute and chronic biliary/cholestatic diseases. Pediatric transplant hepatologists need to possess an in-depth familiarity of acute and chronic management including disease processes, presentations, treatment, and transition of care.

The specific functions which define this EPA include:

1. Knowing and applying the epidemiology, pathophysiology, differential diagnosis, workup, natural history, in managing neonatal cholestasis
2. Knowing and applying the epidemiology, pathophysiology, differential diagnosis, workup, natural history in managing children and adolescents with suspected acute and chronic biliary/cholestatic diseases
3. Knowing the role of liver biopsy, radiologic studies, ERCP, and genetic studies for biliary/cholestatic diseases
4. Interpreting and applying basic/translational and clinical research and current clinical guidelines in treatment of children and adolescents with acute and chronic biliary/cholestatic diseases
5. Recognizing when transplant should be considered for biliary/cholestatic diseases of the neonate, child, and adolescent
6. Educating parents and children on biliary/cholestatic disease including cause, treatment and clinical course
7. Leading, directing, and coordinating care for children/adolescents with biliary/cholestatic diseases within the medical system and the community

Judicious Mapping to Domains of Competence

- [X] Patient Care
- [ ] Medical Knowledge
- [X] Practice-Based Learning and Improvement
- [X] Interpersonal and Communication Skills
- [ ] Professionalism
- [ ] Systems-Based Practice
- [ ] Personal and Professional Development
Competencies Within Each Domain Critical to Entrustment Decisions

| PC 3: | Provide transfer of care that ensures seamless transitions |
| PC 6: | Make informed diagnostic and therapeutic decisions that result in optimal clinical judgment |
| PC 7: | Develop and carry out management plans |
| PBLI 6: | Locate, appraise, and assimilate evidence from scientific studies related to their patient’s health problems |
| PBLI 9: | Participate in the education of patients, families, students, residents, and other health professionals |
| ICS 4: | Work effectively as a member or leader of a health care team or other professional group |

Curricula - Overall Goals and Objectives for Transplant Hepatology EPAs

1. Diagnostic List

During the one-year Advanced Pediatric Transplant Hepatology Program, fellows are expected to develop comprehensive skills in the diagnostic evaluation, treatment and follow up of common and uncommon pediatric liver diseases and pediatric liver transplantation with a focus on:

- Clinical expertise in the diagnosis and management of infants, children, and adolescents with liver diseases
- Indications and contraindications for liver transplantation
- Pre-, peri-, and post-operative management of pediatric liver transplant recipients
- Short- and long-term management of pediatric patients who have undergone liver transplantation

The fellow should become competent in the diagnosis and treatment of the following specific problems:

A. Liver Diseases

- Neonatal cholestasis
- Biliary atresia
- Alagille syndrome
- Progressive familial intrahepatic cholestasis
- Autoimmune hepatitis
- Primary sclerosing cholangitis
- Wilson’s disease
- Nonalcoholic fatty liver disease
- Alpha-1-antitrypsin deficiency
- Viral Hepatitis: Hepatitis A, Hepatitis B, Hepatitis C, Hepatitis E, Non A-E Hepatitis, EBV, CMV
- Acute liver failure
- Drug hepatotoxicity: Including Tylenol induced liver disease, TPN induced liver disease
- Choledochal cyst
- Cirrhosis and portal hypertension
Entrustable Professional Activities
EPA 3 for Pediatric Transplant Hepatology

- Metabolic liver disease
- Impaired growth in children with liver disease and nutritional support of patients with chronic liver disease

B. Liver Transplantation

- Indications for liver transplantation
- Contraindications for liver transplantation (absolute and relative)
- Types of liver transplantation: deceased vs living donor, whole organ vs left lobe vs left lateral vs right lobe
- Multidisciplinary evaluation of liver transplant candidates
- Psychosocial evaluation of liver transplant candidates
- Ethical considerations related to living donation, donation after cardiac death, donation after brain death
- Immunosuppressive medications (indications, complications)
- Acute and chronic complications of liver transplant including:
  - Primary nonfunction
  - Hepatic artery thrombosis
  - Portal vein thrombosis
  - Acute cellular rejection
  - Chronic rejection
  - Biliary problems
  - Infection (EBV, adenovirus, CMV, fungal infection)
  - PTLD
  - Prevention of recurrent viral hepatitis

- Evaluation/indications for emergent re-operation or re-transplantation
- Transplant immunology (including blood group matching, histocompatibility, tissue typing)

2. Procedure List

During the one year of training, the fellow will become proficient in performance and interpretation of the following procedures:

- Percutaneous liver biopsy
- Endoscopy
- Variceal sclerosis/band ligation
- Paracentesis

Proficiency is defined as understanding the clinical indications and contraindications, proper sedation techniques, and intra-procedural monitoring, physical performance of the procedure, interpretation of abnormal and normal findings, and proper post-procedural monitoring.
INPATIENT SERVICE

1. Patient Care

- Fellows must be able to provide patient care that is compassionate, appropriate, and effective for the treatment of health problems and the promotion of health.
- Fellows must acquire the necessary clinical skills to develop expertise in the ability to perform a history and physical examination, make diagnostic and therapeutic decisions, develop and carry out management plans, counsel patients and families, and use information technology to optimize patient care relevant to pediatric hepatology.
- Fellows must have the skills needed to care for patients in all the phases of transplant care including evaluation and indications, pre-transplant management, peri-operative care, immediate postoperative critical care, and the specifics of short-term and long-term post-transplant medical management.
- Fellows must develop competency in managing post-transplant immunosuppression.
- Fellows must develop expertise in diagnosis and management of children with:
  - Chronic cholestasis
  - Cirrhosis
  - End-stage liver disease
  - Acute liver failure
  - Metabolic liver disease
  - Viral hepatitis
  - Autoimmune hepatitis and sclerosing cholangitis
  - Drug hepatotoxicities

- Fellows must understand:
  - The impact of chronic liver disease on growth and development in children
  - Nutritional support of patients with chronic liver disease
  - Problems associated with end stage liver disease including nutritional complications, upper gastrointestinal hemorrhage, refractory ascites, hepatorenal syndrome, and hepatic encephalopathy

- Fellows must develop knowledge of indications and strategies for liver transplantation and recognition of absolute and relative contraindications for liver transplantation including:
  - Psychosocial evaluation of candidates and recipients and their families
  - Primary evaluation, presentation, and discussion of potential liver transplant candidates for consideration by a multidisciplinary board
  - Knowledge of the current UNOS allocation policies
  - Ethical considerations relating to liver transplant donors, including questions related to living donors, donation after cardiac death, criteria for brain death, and appropriate recipients
  - Indications for emergent reoperation or re-transplantation
  - Development of a knowledge base in transplant immunology, including blood group matching, histocompatibility and tissue typing
• Fellows must know how to care for patients that receive technical variant grafts such as living donor grafts, left lobe grafts, left lateral segment grafts.
• Fellows must understand care for transplant recipients including:
  o Peritransplant management (understand different operative phases, warm and cold ischemia time, different vascular and biliary reconstructions, anesthesia issues, fluid and volume management, immediate surgical complications)
  o Prevention and management of opportunistic infection in the transplant recipient (cytomegalovirus, adenovirus, fungal infection, and the spectrum of Epstein-Barr virus related disease including post-transplant lymphoproliferative disease)
  o Prevention and management of recurrent viral hepatitis in the allograft
  o Recognition, evaluation, diagnosis, and treatment of acute and chronic allograft rejection
  o Recognition and intervention for complications of immunosuppressive therapy
  o Recognition, evaluation, and management of long-term complications of liver transplantation
  o Fellows must be able to competently perform and interpret all medical, diagnostic, and surgical procedures considered essential for the area of practice including interpretation of the results of laboratory tests of hepatic function, radiologic imaging (CT, MRI, US), neuropsychological tools for assessment of encephalopathy, and liver biopsies.
  o Fellows must be able to know when to perform allograft biopsies and how to interpret liver transplant biopsy specimens. Fellows must know indications for IR ultrasound guided biopsies.

2. Medical Knowledge
• Fellows must demonstrate knowledge of established and evolving biomedical, clinical, epidemiological, and social-behavioral sciences, as well as application of this to knowledge to patient care.
  o Working knowledge of liver transplantation including the management of patients with end-stage liver disease including nutritional complications, upper GI hemorrhage, refractory ascites, hepatorenal syndrome, hepatic encephalopathy
  o Knowledge of different methods of vascular and biliary reconstruction
  o Knowledge of warm and cold ischemia, anhepatic phase, reperfusion phase
  o Knowledge of organizational principles of a multidisciplinary transplant program including training/responsibilities of nurse coordinators, procurement coordinators, other staff
  o Knowledge of UNOS allocation polices, past and present

• Fellows must go on 3 deceased donor liver procurements to understand principles of donor selection and management.
• Fellows must observe 3 liver transplants.

3. Interpersonal Skills and Communication
• Fellows will learn appropriate supervision of house staff, medical students, and GI fellows involved in direct inpatient care of patients with liver disease and those who are transplant candidates/recipients (supervised by the inpatient or on-call attending).
• The fellow must demonstrate interpersonal and communication skills that result in the effective exchange of written and oral communication with:
Entrustable Professional Activities
EPA 3 for Pediatric Transplant Hepatology

- Patients and families
- Support personnel
- Consultative services

4. Practice-Based Learning

- **Fellow’s Report**: Weekly 30-minute presentations on an organized case history and discussion of laboratory or radiographic findings and therapeutic interventions. These are interactive and involve faculty and fellows.
- **Inpatient Rounds**: Lead by attending/fellow in an interactive discussion

5. Professionalism

- Fellows must demonstrate a commitment to carrying out professional responsibilities and an adherence to ethical principles.
- Fellows are expected to demonstrate professionalism throughout education during physician-patient, physician-family, physicians-physician/allied health professional, and physician-society relationships.
- The fellow will complete all medical records (transplant consults within 24 hours, operative notes within one hour, outpatient notes within 48 hours).

6. Systems-Based Practice

- Fellows must demonstrate an awareness of and responsiveness to the larger context and system of health care, as well as the ability to call effectively on other resources in the system to provide optimal health care.
- Fellows are expected to develop knowledge in topics such as the economics of health care and current health care management issues.
- Fellows must participate in the prevention of medical errors and quarterly M and M’s when a liver/transplant patient is discussed.
- Formal oral communication skills (teaching) are listed under “Medical Knowledge” and include topic lectures and Fellow’s Conferences.
- Fellows will participate in multidisciplinary conferences on a weekly/monthly basis including:
  - Pathology: Lead by faculty liver pathologist, supervised by an attending hepatologist
  - Radiology: Lead by faculty hepatobiliary radiologist, supervised by an attending hepatologist
  - Nutrition: Lead by service nutritionist with faculty supervision on a case-by-case discussion
  - Transplant case presentation (weekly): Hepatologist, surgeon, nurse practitioner, pharmacist, dietician, social workers, psychologist, anesthesiologist, infectious disease physician all present and participate

OUTPATIENT LIVER CLINIC

1. Patient Care

- Fellows must be able to provide patient care that is compassionate, appropriate, and effective for the treatment of health problems and the promotion of health.
• Fellows must acquire the necessary clinical skills to develop expertise in the ability to perform a history and physical examination, make diagnostic and therapeutic decisions, develop and carry out management plans, counsel patients and families, and use information technology to optimize patient care relevant to pediatric hepatology.
• Fellows must develop competency in managing post-transplant immunosuppression.
• Fellows must develop expertise in diagnosis and management of children with:
  o Chronic cholestasis
  o Cirrhosis
  o End-stage liver disease
  o Acute liver failure
  o Metabolic liver disease
  o Viral hepatitis
  o Autoimmune hepatitis and sclerosing cholangitis
  o Drug hepatotoxicities
• Fellows must understand:
  o The impact of chronic liver disease on growth and development in children
  o Nutritional support of patients with chronic liver disease
  o Problems associated with end stage liver disease including nutritional complications, upper gastrointestinal hemorrhage, refractory ascites, hepatorenal syndrome, and hepatic encephalopathy
• Fellows must develop knowledge of indications and strategies for liver transplantation and recognition of absolute and relative contraindications for liver transplantation including:
  o Psychosocial evaluation of candidates and recipients and their families
  o Primary evaluation, presentation, and discussion of potential liver transplant candidates for consideration by a multidisciplinary board
  o Knowledge of the current UNOS allocation policies
  o Ethical considerations relating to liver transplant donors, including questions related to living donors, donation after cardiac death, criteria for brain death, and appropriate recipients
  o Indications for emergent reoperation or re-transplantation
  o Development of a knowledge base in transplant immunology, including blood group matching, histocompatibility and tissue typing
• Fellows must know how to care for patients that receive technical variant grafts such as living donor grafts, left lobe grafts, left lateral segment grafts.
• Fellows must understand care for transplant recipients including:
  o Peritransplant management (understand different operative phases, warm and cold ischemia time, different vascular and biliary reconstructions, anesthesia issues, fluid and volume management, immediate surgical complications)
  o Prevention and management of opportunistic infection in the transplant recipient (cytomegalovirus, adenovirus, fungal infection, and the spectrum of Epstein-Barr virus related disease including post-transplant lymphoproliferative disease)
2. Medical Knowledge

- Fellows must demonstrate knowledge of established and evolving biomedical, clinical, epidemiological, and social-behavioral sciences, as well as application of this to knowledge to patient care.
  
  - Working knowledge of liver transplantation including the management of patients with end-stage liver disease including nutritional complications, upper GI hemorrhage, refractory ascites, hepatorenal syndrome, hepatic encephalopathy
  - Knowledge of different methods of vascular and biliary reconstruction
  - Knowledge of warm and cold ischemia, anhepatic phase, reperfusion phase
  - Knowledge of organizational principles of a multidisciplinary transplant program including training/responsibilities of nurse coordinators, procurement coordinators, other staff
  - Knowledge of UNOS allocation polices, past and present

3. Interpersonal Skills and Communication

- The fellow must demonstrate interpersonal and communication skills that result in the effective exchange of written and oral communication with:
  
  - Patients and families
  - Support personnel
  - Consultative services

- The fellow will learn to participate in a multidisciplinary transplant team (including hepatologists, surgeons, nurse coordinators, dieticians, pharmacists, social workers, anesthesiologists, infectious disease physicians).

4. Practice-Based Learning

- **Fellow’s Report:** Weekly 30-minute presentations on an organized case history and discussion of laboratory or radiographic findings and therapeutic interventions. These are interactive and involve faculty and fellows
- **M and M:** Quarterly
5. Professionalism

- Fellows must demonstrate a commitment to carrying out professional responsibilities and an adherence to ethical principles.
- Fellows are expected to demonstrate professionalism throughout education during physician-patient, physician-family, physicians-physician/allied health professional, and physician-society relationships.
- The fellow will complete all medical records (transplant consults within 24 hours, operative notes within one hour, outpatient notes within 48 hours).

6. Systems-Based Practice

- Fellows must demonstrate an awareness of and responsiveness to the larger context and system of health care, as well as the ability to call effectively on other resources in the system to provide optimal health care.
- Fellows are expected to develop knowledge in topics such as the economics of health care and current health care management issues.
- Fellows must participate in the prevention of medical errors and quarterly M and M’s when a liver/transplant patient is discussed.
- Formal oral communication skills (teaching) are listed under “Medical Knowledge” and include topic lectures and Fellow’s Conferences.
- Fellows will participate in multidisciplinary conferences on a weekly/monthly basis including:
  - Pathology: Lead by faculty liver pathologist, supervised by an attending hepatologist
  - Radiology: Lead by faculty hepatobiliary radiologist, supervised by an attending hepatologist
  - Nutrition: Lead by service nutritionist with faculty supervision on a case-by-case discussion
  - Transplant case presentation (weekly): Hepatologist, surgeon, nurse practitioner, pharmacist, dietician, social workers, psychologist, anesthesiologist, infectious disease physician all present and participate

PROCEDURE CENTER

1. Patient Care

- Fellows must perform at least 15 liver biopsies.
- Fellows must be able to perform upper endoscopy with variceal banding/sclerotherapy.

2. Medical Knowledge

- Fellows must be able to know when to perform allograft biopsies and how to interpret liver transplant biopsy specimens. Fellows must know indications for IR ultrasound guided biopsies.

3. Interpersonal Skills and Communication

- The fellow must demonstrate interpersonal and communication skills that result in the effective exchange of information.
4. Practice-Based Learning
   • M and M: Quarterly

5. Professionalism
   • Fellows are expected to demonstrate professionalism throughout education during physician-patient, physician-family, physicians-physician/allied health professional, and physician-society relationships.
   • The fellow will complete all medical records (procedure notes within one hour of procedure).

6. System-Based Practice
   • Fellows must demonstrate an awareness of and responsiveness to the larger context and system of health care, as well as the ability to call effectively on other resources in the system to provide optimal health care.
   • Fellows are expected to develop knowledge in topics such as the economics of health care and current health care management issues.

ADULT HEPATOLOGY ROTATION

1. Patient Care
   • Fellows must develop knowledge of indications and strategies for liver transplantation and recognition of absolute and relative contraindications for liver transplantation including:
     o Psychosocial evaluation of candidates and recipients and their families
     o Primary evaluation, presentation, and discussion of potential liver transplant candidates for consideration by a multidisciplinary board
     o Knowledge of the current UNOS allocation policies
     o Ethical considerations relating to liver transplant donors, including questions related to living donors, donation after cardiac death, criteria for brain death, and appropriate recipients

2. Medical Knowledge
   • Fellows must observe/participate in one living donation evaluation and procedure.

3. Interpersonal Skills and Communication
   • The fellow must demonstrate interpersonal and communication skills that result in the effective exchange of information.

4. Practice-Based Learning
   • Participate in any M and M’s involved in living donation

5. Professionalism
   • Fellows must demonstrate a commitment to carrying out professional responsibilities and an adherence to ethical principles.
6. Systems-Based Practice

- Fellows must demonstrate an awareness of and responsiveness to the larger context and system of health care, as well as the ability to call effectively on other resources in the system to provide optimal health care.

Transplant Hepatology Authors

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