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Evaluation and Management of a Patient with a Hepatobiliary Disorder

Description of the Activity	Biliary atresia (BA) and choledochal cyst (CC) are congenital diseases that occur in infancy and early childhood. Infants with BA and CC are frequently referred to pediatric general surgeons in the inpatient and outpatient settings. The pediatric surgeon should be able to perform and counsel families on fetal risk stratification and diagnose and treat patients with these conditions, selecting the optimal operative treatment, approach, and timing. Patient- and family-centered, evidenced-based, and shared decision-making is necessary to ensure ideal outcomes.
Functions	❖ Nonoperative/Preoperative ❖ BA ➢ Obtain and synthesize essential information from the patient's pre- and postnatal records, history, physical examination, and initial diagnostic evaluations to develop a differential diagnosis. Correlate this information with any known prenatal imaging. ➢ Evaluate a patient with neonatal jaundice, investigating for possible congenital, genetic, infectious, and metabolic causes. ➢ Obtain the information needed to diagnose BA, including laboratory (blood studies), radiologic (cholangiogram, hepatobiliary iminodiacetic acid scan, ultrasound), and surgical options (liver biopsy, cholangiogram). ➢ Demonstrate understanding of other conditions that can mimic BA, such as hepatitis, alpha-1 antitrypsin disorder, and Alagille syndrome, and diagnose these conditions. ➢ Complete a cost-effective, evidence-based diagnostic evaluation, obtaining appropriate imaging directed at operative planning. ➢ Identify the optimal timing, approach, and technique for repair based on the patient's diagnosis and clinical stability (timing of a portoenterostomy, need for preoperative antibiotics and vitamin K, need for cholangiogram and portoenterostomy). ➢ Recognize when a patient with delayed presentation or advanced cirrhosis should be referred to the transplant team. ➢ Communicate the diagnosis and treatment options to the family and consultants. ➢ Obtain informed consent, describing the indications, risks, benefits, alternatives, and potential complications of the planned operation, including nuances relevant to the patient's individual condition and comorbidities, and ensure familial understanding. Document the informed consent discussion in the medical record. ➢ Devise an operative plan, and communicate it to the operative team (anesthesia, nursing, techs, assistants), including patient position, anesthesia needs, special instrumentation, and postoperative planning. ❖ CC ➢ Obtain and synthesize essential information from the patient's pre- and postnatal records, history, physical examination, and initial diagnostic evaluations to develop a differential diagnosis. Correlate this information with any known prenatal imaging. ➢ Evaluate a patient with neonatal jaundice, investigating for possible congenital, genetic, infectious, mechanical and metabolic causes. ➢ Demonstrate knowledge of the 5 types of CC. ➢ Complete a cost-effective, evidence-based diagnostic evaluation, obtaining imaging directed at operative planning (ultrasound, magnetic resonance cholangiopancreatography [MRCP], endoscopy) and assessing the need for preoperative antibiotics. ➢ Identify the optimal timing, approach, and technique for repair based on the patient's diagnosis and clinical stability.

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- Determine which patients require intervention in the neonatal period and which are stable enough to undergo elective operative intervention.
- Determine when a patient would benefit from a minimally invasive versus an open approach.

➢ Identify the appropriate surgical procedure for each of the 5 types of CC, including hepaticoduodenostomy and hepaticojejunostomy, endoscopic treatment, and drain placement.

➢ Communicate the diagnosis and treatment options to the family and consultants.

➢ Obtain informed consent, describing the indications, risks, benefits, alternatives, and potential complications of the planned operation, including nuances relevant to the patient's individual condition and comorbidities, and ensure familial understanding. Document the informed consent discussion in the medical record.

➢ Devise an operative plan, and communicate it to the operative team (anesthesia, nursing, techs, assistants), including patient position, anesthesia needs, special instrumentation, and postoperative planning.

❖ Intraoperative

❖ BA

➢ Manage the perioperative environment, including room setup, equipment check, preprocedural time-out, specimen processing, counts, wound classification, updating the family, and debriefing functions.

➢ Position the patient to expose the operative field, taking precautionary measures to avoid iatrogenic injury and allow the anesthesia team access to the patient.

➢ Confirm the availability of necessary equipment (staplers, energy devices, clip appliers, ties, extra equipment).

➢ Collaborate with other health care professionals to create and maintain an intraoperative environment that promotes safe patient care.

➢ Perform the procedures required to manage BA through an open approach, including portoenterostomy.

➢ Recognize and develop a management plan for unexpected intraoperative findings, such as aberrant anatomy, bleeding liver and hepatic vessels, and bile leak.

❖ CC

➢ Manage the perioperative environment, including room setup, equipment check, preprocedural time-out, specimen processing, counts, wound classification, updating the family, and debriefing functions.

➢ Position the patient to expose the operative field, taking precautionary measures to avoid iatrogenic injury and allow the anesthesia team access to the patient.

➢ Confirm the availability of necessary equipment (staplers, energy devices, clip appliers, ties, extra equipment).

➢ Collaborate with other health care professionals to create and maintain an intraoperative environment that promotes safe patient care.

➢ Perform the procedures required to manage CC through minimally invasive and open approaches, including hepaticoduodenostomy and hepaticojejunostomy.

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	➢ Recognize and develop a management plan for unexpected intraoperative findings, including aberrant anatomy; bleeding from hepatic arteries/veins, portal vein, and IVC; bile leak; and significant inflammation involving the CC and portal vein. ➢ Recognize indications to convert from a minimally invasive to an open approach ➢ Identify the indications for endoscopic retrograde cholangiopancreatography (ERCP). ❖ Postoperative • BA ○ Provide postoperative management for a patient with BA. ○ Communicate with the family/caregiver(s) and the health care team to ensure instructions are understood. ○ Establish goals for drain removal if used. ○ Identify early (anastomotic leak, ileus, cholangitis) and late (liver failure requiring transplant, bowel obstruction) postoperative complications associated with a portoenterostomy procedure. ○ Recognize the long-term outcomes from portoenterostomy, such as possible cholangitis and the fact that 30% will avoid transplantation, 30% will have liver failure in the first year, and 30% will progress to liver failure later in childhood. ○ Recognize the need for a transplant referral after portoenterostomy in patients with early failed portoenterostomy, repeated cholangitis, and cirrhosis. ○ Demonstrate understanding of evidence-based best practices, including avoidance of the routine use of steroids pre- or postoperatively ○ Plan and establish a short- and long-term follow-up plan that includes pathology and imaging as needed. • CC ○ Provide postoperative management for a patient with CC. ○ Communicate with the family/caregiver(s) and the health care team to ensure instructions are understood. ○ Establish goals for drain removal if used. ○ Identify early (anastomotic leak, ileus, bleeding) and late (bowel obstruction and stricture) postoperative complications associated with a portoenterostomy procedure. ○ Plan and establish a short- and long-term follow-up plan that includes pathology and imaging as needed. Perform postoperative care, including administration of antibiotics, and recognize the acute surgical complications that are associated with surgical treatment, such as ileus.
Scope	❖ In scope • Diagnoses ○ BA ○ CC ○ Workup for neonatal jaundice: ▪ Congenital ▪ Genetic ▪ Infectious

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- Mechanical
- Metabolic

- Procedures
  - Hepaticojejunostomy
  - Intraoperative cholangiography
  - Laparoscopic versus open choledochal cyst resection with hepaticoduodenostomy
  - Liver biopsy
  - Portoenterostomy
  -
- Special populations
  - Prenatally diagnosed cystic BA
  - Type 4 choledochal cysts
  - Inflamed choledochal cyst that cannot be separated

❖ Out of scope

- Diagnoses/procedures
  - Bile duct cancer
  - Gallbladder cancer
  - Hepatic angiosarcoma
  - Hepatoblastoma
  - Neonatal vascular malformation and vascular tumor

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Level	Nonoperative/Preoperative	Intraoperative	Postoperative
1 **Framework:** The attending will show and tell or the learner acts as first assistant. **Entrustment:** The learner demonstrates understanding of information and has basic skills. What a new pediatric surgery fellow should know.	• With active guidance, obtains and synthesizes essential information from a patient's pre- and postnatal records, H&P, initial diagnostic evaluation, and prenatal imaging to develop a differential, including the type of biliary atresia/choledochal cyst • With active guidance, defines the appropriate operative timing, approach, and technique based on patient diagnosis and communicates these to the patient and family • With active guidance, identifies the different surgical procedures utilized in patients with biliary atresia/choledochal cyst (portoenterostomy, hepaticoduodenostomy, hepaticojejunostomy, endoscopic treatment, drain placement) • During the initial evaluation, establishes professional rapport with family and consultants and respectfully relays the pertinent clinical findings in a clear and understandable manner • Establishes a therapeutic relationship with a patient and family and has a basic understanding of how personal biases can lead to communication barriers	• Demonstrates basic knowledge of biliary atresia/choledochal cyst and anatomic variations • Recognizes the specific functions of the periop environment, including room setup, equipment check, time-out, and debrief • With active guidance, progresses through the initial portions of liver mobilization or cyst dissection, including basic interpretation of the cholangiogram • Demonstrates basic understanding of intraop complications due to aberrant anatomy such as bleeding or bile leak	• With active guidance, implements a postop management plan for a patient with uncomplicated biliary atresia or choledochal cyst • Demonstrates basic understanding of acute surgical complications associated with surgical treatment of biliary atresia or choledochal cyst (eg, anastomotic leak, ileus, bleeding, cholangitis, liver failure, bowel obstruction/stricture) • With active guidance, follows an evidence-based long-term management plan for a patient with biliary atresia or choledochal cyst • Engages family and care team in discussion of postoperative instructions and respectfully relays information on goals, and expectations to • With active guidance, uses active listening to adapt communication style to fit family/guardians/care team needs; coordinates recommendations from different members of the health care team to optimize patient care; and maintains effective communication in a crisis situation • Respectfully listens to concerns from members of care teams

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Level	Nonoperative/Preoperative	Intraoperative	Postoperative
2 **Framework:** The learner demonstrates understanding of the steps of the operation but requires direction through principles and does not know the nuances of a basic case. **Entrustment:** The learner can use the tools but may not know exactly what, where, or how to do it. The attending gives active help throughout the case to maintain forward progression or may need to take over the case at a certain point	• With direct supervision, obtains and synthesizes essential information from a patient's pre- and postnatal records, H&P, initial diagnostic evaluation and prenatal imaging to develop a differential, including the type of biliary atresia/choledochal cyst • With direct supervision, defines the appropriate operative timing, approach, and technique based on patient diagnosis and communicates these to the patient and family • With direct supervision, identifies the appropriate surgical procedure for each of the types of biliary atresia/choledochal cyst (portoenterostomy, hepaticoduodenostomy, hepaticojejunostomy, endoscopic treatment, drain placement) • With direct supervision, communicates the diagnosis and treatment options to a family and consultants and obtains informed consent • Establishes a therapeutic relationship with a patient and family and when prompted, reflects on personal biases while attempting to minimize communication barriers	• With direct supervision, identifies surgically relevant anatomic variations with an intraop cholangiogram, performs liver biopsy, and alters patient management appropriately • With direct supervision, manages the periop environment, including room setup, equipment check, time-out, and debrief • With direct supervision, progresses through the initial portions of liver mobilization, anterior cyst dissection for choledochal cyst, and hepaticoduodenostomy/ hepaticojejunostomy anastomosis; assists with portal plate dissection and portoenterostomy for biliary atresia • With direct supervision, manages, anticipates, and prevents intraop complications for portoenterostomy, hepaticoduodenostomy, or hepaticojejunostomy anastomosis	• With direct supervision, manages the postop course of a patient with uncomplicated biliary atresia or choledochal cyst • With direct supervision, identifies and manages surgical complications commonly associated with surgical treatment of biliary atresia or choledochal cyst (eg, anastomotic leak, ileus, bleeding, cholangitis, liver failure, bowel obstruction/stricture) • With passive guidance, follows an evidence-based long-term management plan for a patient with biliary atresia or choledochal cyst • With direct supervision, communicates postop instructions, goals, and expectations to family/guardians/care teams clearly and concisely in an organized and timely manner • Under direct supervision, uses active listening to adapt communication style to fit family/guardians/care team needs; coordinates recommendations from different members of the health care team to optimize patient care; and maintains effective communication in a crisis situation • Under direct supervision, communicates feedback to and

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Level	Nonoperative/Preoperative	Intraoperative	Postoperative
			requests constructive criticism from superiors
3 **Framework:** The learner has a good understanding of surgical options and techniques but does not recognize abnormalities and does not understand the nuances of a complicated case. **Entrustment:** The learner can perform the operation/task independently in the uncomplicated patient or The attending provides passive/indirect supervision/suggestions in the complicated patient but still allows the learner to perform the operation/task themselves.	• With indirect supervision, integrates essential information from a patient's pre- and postnatal records, H&P, initial diagnostic evaluation and prenatal imaging to develop a treatment plan for patients with biliary atresia/choledochal cyst • With indirect supervision, defines the appropriate operative timing, approach, and technique based on patient diagnosis and participates in shared decision-making to align the values, goals, and preferences of the patient and family with treatment options to make a personalized care plan • With indirect supervision, identifies the appropriate surgical procedure for each of the types of biliary atresia/choledochal cyst (portoenterostomy, hepaticoduodenostomy, hepaticojejunostomy, endoscopic treatment, drain placement) • With indirect supervision, communicates the diagnosis and treatment options to a family and consultants and obtains informed consent • Establishes a therapeutic relationship with a patient and family and with minimal prompting critically reflects on personal biases while attempting to minimize communication barriers	• With indirect supervision, identifies surgically relevant anatomic variations with an intraop cholangiogram, performs liver biopsy, and alters patient management appropriately • With indirect supervision, manages the periop environment, including room setup, equipment check, time-out, and debrief • With indirect supervision, progresses through liver mobilization, circumferential cyst dissection for choledochal cyst, and hepaticoduodenostomy/ hepaticojejunostomy anastomosis; assists with portal plate dissection and portoenterostomy for biliary atresia • With indirect supervision, manages, anticipates, and prevents intraop complications due to aberrant anatomy such as bleeding and bile leak	• With indirect supervision, implements a postop management plan for a patient with complicated biliary atresia or choledochal cyst • With indirect supervision, recognizes and manages surgical complications commonly associated with surgical treatment of biliary atresia or choledochal cyst (eg, anastomotic leak, ileus, bleeding, cholangitis, liver failure, bowel obstruction/stricture) • With indirect supervision, integrates patient- and family-specific factors into the construction of an evidence-based long-term management plan for a patient with biliary atresia or choledochal cyst, including the use of steroids in biliary atresia, and relays outcomes to the family • With indirect supervision, ensures that postop instructions, goals, and expectations are understood by family/guardians/care teams • Often uses active listening to adapt communication style to fit family/guardians/care team needs; coordinates recommendations from different members of the health care team to optimize patient care; and

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Level	Nonoperative/Preoperative	Intraoperative	Postoperative
			maintains effective communication in a crisis situation • With indirect supervision, communicates feedback to and requests constructive criticism from superiors
4 **Framework** The learner has a strong and indepth understanding of surgical options and techniques. **Entrustment:** Can perform the operation/task independently in complicated cases or The attending may need to provide indirect supervision or suggestions in the context of extremely rare or severely complicated cases	• Independently integrates and synthesizes essential information from a patient's pre- and postnatal records, H&P, initial diagnostic evaluation and prenatal imaging to develop a treatment plan for patients with biliary atresia/choledochal cyst • Independently defines and implements the appropriate operative timing, approach, and technique based on patient diagnosis and participates in shared decision-making to align the values, goals, and preferences of the patient and family with treatment options to make a personalized care plan • Independently identifies the appropriate surgical procedure for each of the types of biliary atresia/choledochal cyst (portoenterostomy, hepaticoduodenostomy, hepaticojejunostomy, endoscopic treatment, drain placement) • Independently communicates the diagnosis and treatment options to a family and consultants and obtains informed consent • Independently recognizes personal biases while attempting to proactively minimize	• Independently demonstrates comprehensive knowledge of biliary atresia/choledochal cyst and anatomic variations such as surgically correctable types of biliary atresia or aberrant hepatic arterial blood supply • Independently manages the periop environment, including room setup, equipment check, time-out, and debrief • Independently progresses fluidly through the entirety of portoenterostomy (including cholangiogram and liver biopsy) or circumferential cyst resection and hepaticoduodenostomy or hepaticojejunostomy anastomosis • Independently manages, anticipates, and prevents intraop complications due to aberrant anatomy such as bleeding, bile leak, and adherent posterior wall	• Independently implements a postop management plan for a patient with biliary atresia or choledochal cyst • Independently recognizes and manages surgical complications commonly associated with surgical treatment of biliary atresia or choledochal cyst (eg, anastomotic leak, ileus, bleeding, cholangitis, liver failure, bowel obstruction/stricture) • Independently integrates patient- and family-specific factors into the construction of an evidence-based long-term management plan for a patient with biliary atresia or choledochal cyst, including the use of steroids in biliary atresia, and relays outcomes to the family • Independently communicates with family/guardians/care teams to ensure postop instructions, goals, and expectations are understood • Consistently uses active listening to adapt communication style to fit family/guardians/care team needs;

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Level	Nonoperative/Preoperative	Intraoperative	Postoperative
	communication barriers with family/caregivers/health care teams		coordinates recommendations from different members of the health care team to optimize patient care; and maintains effective communication in a crisis situation • Consistently and independently communicates feedback to and requests constructive criticism from superiors