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Evaluation and Management of a Patient with a Congenital Diaphragmatic Hernia

Description of the Activity	Congenital diaphragmatic hernia (CDH) is a cornerstone diagnosis in pediatric surgery. Although rare, it is associated with significant individual and societal costs. The care of a patient with CDH may start with the prenatal diagnosis and continue through adolescence. The essential functions of this activity include prenatal counseling, complex critical care including extracorporeal life support (ECLS), definitive repair, and surveillance. The spectrum of disease includes Morgagni and Bochdalek hernias as well as diaphragmatic eventration.
Functions	❖ Nonoperative/Preoperative • Perform a prenatal workup and provide counseling accordingly. • Recognize when transfer is indicated. • Provide preoperative critical care, including ventilator management and ECLS. • Determine the timing of repair. • Recognize potential complications related to treatment. • Differentiate CDH from diaphragmatic eventration, and manage diaphragmatic eventration. • Recognize the signs and symptoms of delayed presentation. • Recognize CDH on prenatal ultrasound. • Perform risk stratification using prenatal ultrasound and magnetic resonance imaging to describe the defect side, lung/head ratio, observed-to-expected lung/head ratio, liver and stomach position, fetal lung volume, and cardiac assessment, and use this information to place the patient into high-, moderate-, or low-risk categories. • Perform prenatal counseling, taking into account concomitant anomalies, such as the likelihood of survival, likelihood/candidacy of extracorporeal membrane oxygenation (ECMO), expected hospital course, long-term outcomes, and the delivery plan. • Recognize when a patient is a candidate for fetoscopic endotracheal occlusion (FETO). • Recognize the need for a multidisciplinary team to ensure proper prenatal care, surveillance, and counseling as well as coordination of delivery location. • Identify ventilator strategies with permissive hypercapnia, recognizing the importance of targeted strategies in early management. • Identify the need for adjunct studies upon diagnosis. • Recognize the need for ECLS and contraindications to ECLS. • Consider concomitant anomaly management. • 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 positioning, anesthesia needs, special instrumentation, and postoperative planning. • Recognize the implications of repair while the patient is on ECLS, including timing (early vs late) and location (neonatal intensive care unit, operating room) and their associated advantages/disadvantages. • Determine the need for surgical repair of eventration.

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	❖ Intraoperative
	• Perform procedures as indicated:
	  ○ Open, thoracoscopic, or laparoscopic repair
	  ○ Primary, muscle flap, or mesh repair
	  ○ Repair on ECLS
	• Systematically examine the defect.
	• Identify the sac if present.
	• Repair the defect.
	• Pay specific attention to the posterolateral portion, and use rib sutures if necessary.
	• Determine if an open abdominal incision can be closed primarily, and perform alternative closures if necessary.
	• Recognize the treatment plan when a sequestration is identified intraoperatively.
	• Perform plication in a patient with eventration.
	
	❖ Postoperative
	• Communicate the postoperative plan of care to the family and other involved health care team members.
	• Perform critical care of the patient, including weaning to extubation, weaning from the ECLS circuit if used, starting feeds, monitoring for compartment syndrome, managing an open abdomen if present, and managing other complications such as hemorrhage and chylothorax.
	• Monitor the patient for early recurrence.
	• Perform outpatient surveillance for delayed recurrence.
	• Identify long-term morbidity (chronic lung disease, gastroesophageal reflux, failure to thrive, neurocognitive dysfunction).
	• Recognize the importance of a multidisciplinary approach to long-term follow-up.
	• Identify the implications of anatomic variance in intestinal rotation.
	• Provide postoperative critical care.
	• Provide postoperative surveillance after discharge.

Scope	❖ In scope
	
	• Diagnoses
	  ○ Congenital diaphragmatic hernia:
	  ▪ Bochdalek
	  ▪ Morgagni
	• Diaphragmatic eventration
	
	• Procedures
	  ○ Laparoscopic repair

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	○ Muscle flap ○ Open CDH repair +/– mesh ○ Thoracoscopic CDH repair +/– mesh • Special populations ○ Prenatal consultation ○ Patients needing repair on ECLS ❖ Out of scope • Diagnoses/procedures ○ Hiatal hernia ○ Traumatic diaphragmatic hernia

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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 assistance, participates in prenatal consultation, including review of abnormal prenatal radiographic findings and risk stratification • With active guidance, articulates the importance of optimal ventilatory strategies • With active guidance, explains a diagnosis to the multidisciplinary team and family and obtains informed consent • With active guidance, participates in conversations about surgical options, timing of repair, and ECLS strategies • With active guidance, demonstrates understanding of the implications of comorbidities such as congenital heart disease in a low-risk patient • With active guidance, diagnoses and manages diaphragmatic eventration and Morgagni hernia • Demonstrates how to access and use available evidence for preop management and incorporates the patient's and family's preferences and values into care • With active guidance, explains the diagnosis to the multidisciplinary team and family	• Requires active assistance in surgical positioning and prep • With active guidance, demonstrates basic understanding of key steps of the operation, including examining the defect, identifying the sac if present, and repairing the defect, needing substantial guidance when choosing the type of procedure (open vs thoracoscopic vs lap) and repair (primary vs muscle flap vs mesh, rib sutures) • Requires active guidance to manage concurrent sequestration • Requires active guidance to perform diaphragm plication for eventration • With active guidance, can repair a diaphragmatic hernia recurrence • Requires active guidance to determine if the abdomen should be closed	• With active guidance, diagnoses postop complications such as compartment syndrome, hemorrhage, and chylothorax, requiring active assistance to manage them • With active guidance, identifies long-term morbidity, including chronic lung disease, reflux, failure to thrive, and neurocognitive dysfunction, recognizing the importance of surveillance for delayed recurrence through a multidisciplinary approach • With active guidance, communicates the basic steps of the operative procedure and the postop plan/expected course with the multidisciplinary team and family, including ventilation management, pulmonary HTN, and nutrition • With active guidance, participates with the ICU team in ventilation and ECLS weaning as necessary • Demonstrates how to access and use available evidence for postop care, integrating it with the patient's and family's preferences and values

THE AMERICAN BOARD OF SURGERY

Evaluation & Management of a Patient with a Congenital Diaphragmatic Hernia

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	• Requires active guidance to diagnose a diaphragmatic hernia recurrence		
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.	• Requires direct supervision during prenatal consultation to review abnormal prenatal radiographic findings and describe risk stratification • With direct supervision, optimizes ventilatory strategies • With direct supervision, explains a diagnosis to the multidisciplinary team and family and obtains informed consent • With direct supervision, provides preoperative planning regarding surgical options, timing of repair, and ECLS strategies • With passive assistance, demonstrates a good understanding of the implications of comorbidities such as congenital heart disease and adjusts treatment accordingly in a low-risk patient • With direct supervision, diagnoses and manages diaphragmatic eventration and Morgagni hernia • Articulates clinical questions about preop management and elicits the patient's and family's preferences and values to guide evidence-based care	• Requires direct supervision for surgical positioning and prep • With direct supervision, performs key steps of the operation, including examining the defect, identifying the sac if present, and repairing the defect; for straightforward cases, chooses the type of procedure (open vs thoracoscopic vs lap) and repair (primary vs muscle flap vs mesh, rib sutures) • Requires direct supervision to identify and manage concurrent sequestration • Requires direct supervision to perform diaphragm plication for eventration • With direct supervision, can repair a diaphragmatic hernia recurrence • Requires passive guidance to determine if the abdomen should be closed	• Diagnoses postop complications such as compartment syndrome, hemorrhage, and chylothorax, requiring direct supervision to manage them • With direct supervision, identifies long-term morbidity, including chronic lung disease, reflux, failure to thrive, neurocognitive dysfunction, and surveillance for delayed recurrence, recognizing the importance of a multidisciplinary approach • With indirect supervision, communicates most aspects of the operative procedure and postop plan/expected course with the multidisciplinary team and family, including ventilation management, pulmonary HTN, and nutrition • With passive guidance, works with the ICU team in ventilation and ECLS weaning as necessary • Articulates clinical questions about postop care and elicits the patient's and family's preferences and values to guide evidence-based care

THE AMERICAN BOARD OF SURGERY

Evaluation & Management of a Patient with a Congenital Diaphragmatic Hernia

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	• With direct supervision, explains the diagnosis to the multidisciplinary team and family • With direct supervision, diagnoses a diaphragmatic hernia recurrence		
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.	• Requires indirect supervision during prenatal consultation to review abnormal prenatal radiographic findings and describe risk stratification • With indirect supervision, optimizes ventilatory strategies • With indirect supervision, explains a diagnosis to the multidisciplinary team and family and obtains informed consent • With indirect supervision, develops a plan regarding surgical options, timing of repair, and ECLS strategies • With passive assistance, demonstrates understanding of the implications of comorbidities such as congenital heart disease and adjusts treatment accordingly in a high-risk patient • With indirect supervision, diagnoses and manages diaphragmatic eventration and Morgagni hernia • Applies the best available evidence to preop management and integrates it with the patient's and family's preferences to guide care	• Requires indirect supervision for surgical positioning and prep • Independently performs key steps of the operation for less severe types of diaphragmatic hernias (A and B); with indirect supervision, performs key steps of the operation for more severe types (C and D), including examining the defect, identifying the sac if present, and repairing the defect; for complex cases, chooses the type of procedure (open vs thoracoscopic vs lap) and repair (primary vs muscle flap vs mesh, rib sutures) • Requires indirect supervision to identify and manage concurrent sequestration • Requires indirect supervision to perform diaphragm plication for eventration • With indirect supervision, can repair a diaphragmatic hernia recurrence • Requires passive guidance to determine when the abdomen needs to be left open, manages the open abdomen, and determines when closure is appropriate	• With indirect supervision, anticipates and implements strategies to manage postop complications such as compartment syndrome, hemorrhage, and chylothorax • With indirect supervision, identifies, counsels, and manage long-term morbidity, including chronic lung disease, reflux, failure to thrive, neurocognitive dysfunction, and surveillance for delayed recurrence, recognizing the importance of a multidisciplinary approach • With indirect supervision, communicates all aspects of the operative procedure and the comprehensive postop plan/expected course with the multidisciplinary team and family, including ventilation management, pulmonary HTN, and nutrition • With indirect supervision, works with the ICU team to individualize ventilation and ECLS management as necessary • Locates and applies the best available evidence for postop care, integrated

THE AMERICAN BOARD OF SURGERY

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	• With indirect supervision, explains the diagnosis to the multidisciplinary team and family and obtains informed consent • With indirect supervision, diagnoses a diaphragmatic hernia recurrence		with the patient's and family's preferences
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 performs a prenatal consult, including imaging interpretation, risk stratification, and counseling • Independently uses common ventilator strategies with permissive hypercapnia to optimize oxygenation/ventilation and determines the need for ECMO • Independently holds a multidisciplinary team/family meeting and obtains informed consent • Independently develops an operative plan regarding surgical options, timing of repair, and ECLS strategies • Independently recognizes relevant comorbidities (congenital heart disease, sequestration) and incorporates these into operative planning in a high-risk patient • Independently diagnoses and manages diaphragmatic eventration and Morgagni hernia	• Independently performs surgical positioning and prep • Independently chooses the best procedure to perform diaphragmatic repair in complex patients (including repair on ECMO and C and D defects) by examining the defect, identifying the sac if present, and repairing the defect • Independently recognizes and manages concurrent sequestration • Independently performs diaphragm plication for eventration • Can independently repair a diaphragmatic hernia recurrence • Independently recognizes when the abdomen needs to be left open, manages the open abdomen, and determines when closure is appropriate	• Independently anticipates and implements strategies to manage postop complications such as compartment syndrome, hemorrhage, and chylothorax • Independently identifies, counsels and manages long-term morbidity, including chronic lung disease, reflux, failure to thrive, neurocognitive dysfunction, and surveillance for delayed recurrence, recognizing the importance of a multidisciplinary approach • Independently communicates all aspects of the operative procedure and the comprehensive postop plan/expected course with the multidisciplinary team and family, including ventilation management, pulmonary HTN, nutrition and customizes emotionally difficult news (eg. changes to the operative plan, adverse outcomes, expectations, additional procedures) to the parent(s)family in a culturally caring manner

THE AMERICAN BOARD OF SURGERY

Evaluation & Management of a Patient with a Congenital Diaphragmatic Hernia

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	• Critically appraises and applies evidence for preop management, even in the face of uncertain or conflicting evidence, to guide care, tailoring it to the patient and family • Independently explains the diagnosis to the multidisciplinary team and family and obtains informed consent • Independently diagnoses a diaphragmatic hernia recurrence		• Actively and independently works with the ICU team to individualize ventilation and ECLS management as necessary • Critically appraises and applies evidence to guide postop care, even in the face of uncertain or conflicting evidence, tailoring it to the patient and family