THE AMERICAN BOARD OF SURGERY

Evaluation and Management of a Patient with an Abdominal Wall Defect

Description of the Activity	Abdominal wall defects are a common congenital anomaly treated by pediatric surgeons that almost always require surgical repair either in the urgent setting immediately after birth or in a delayed fashion. The essential function of this activity is the definitive diagnosis, workup, and treatment of the spectrum of abdominal wall defects from the prenatal phase of care to long-term follow-up.
Functions	❖ Nonoperative/Preoperative • Gastroschisis (GS) ○ Counsel a family on pre- and postnatal strategies to optimize the outcome for the fetus, including the risk of intrauterine fetal demise. ○ Discuss options for surgical repair after birth and the long-term outcomes and risks of GS with the family. ○ Identify risk factors for a fetus with GS, including gastrointestinal dilation, worsening biophysical profile, and intrauterine growth restriction. ○ Identify the risks and benefits of both early and near-term delivery in a fetus with GS. ○ Perform an initial evaluation of the bowel to determine the urgency and type of surgical repair, including the identification of strategies to improve perfusion to the bowel if needed. ○ Discuss the different strategies for GS repair, including the associated risks, benefits, and outcomes of each strategy. ○ Recognize the different types of silos, including prefabricated and surgeon-constructed versions. ○ Identify the need for early gastric decompression, intravenous access, and fluid resuscitation. ○ Recognize the importance of bowel coverage, temperature regulation, patient position, and frequent reassessment of bowel viability. ○ 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. • Omphalocele ○ Counsel the family on the long-term outcomes and risks associated with omphalocele. ○ Identify a safe delivery strategy in a fetus with a giant omphalocele. ○ Perform additional studies to diagnose associated anatomic and genetic anomalies in a fetus with omphalocele. ○ Discuss the spectrum of associated anatomic abnormalities and prenatal diagnostic studies. ○ Perform an initial evaluation of the abdominal wall defect, and identify the difference between an omphalocele and a giant omphalocele based on physical examination. ○ Order testing to allow for safe upfront surgical repair (eg, echocardiogram, postnatal glucose monitoring). ○ Discuss options for a safe surgical plan based on examination of the omphalocele, including the indications for primary omphalocele repair and delayed repair. ○ Formulate an operative plan based on the size of the omphalocele, the severity of comorbid conditions, and the epithelialization of the sac.

Evaluation and Management of a Patient with an Abdominal Wall Defect

- Determine the timing of a delayed ventral hernia repair when using the "paint and wait" strategy for a giant omphalocele.
- Obtain informed consent, describing the indications, risks, benefits, alternatives, and potential complications of the planned operation, including nuances relevant to the patient's condition and comorbidities, and ensure familial understanding. Document the informed consent discussion in the medical record.
- Devise an operative plan, and communicate it to members of the operative team (anesthesia, nursing, techs, assistants), including patient position, anesthesia needs, special instrumentation, and postoperative planning.

- Cloacal exstrophy
  - Perform a focused initial physical examination to identify anatomic variations in the size of the omphalocele and bladder plates.
  - Ensure adequate protective covering of the omphalocele and mucosal surfaces.
  - Order preoperative diagnostic studies and tests (including karyotype) to evaluate for associated abnormalities, including spinal dysraphism (omphalocele, exstrophy of the cloaca, imperforate anus, and spinal defects [OEIS]).
  - Discuss different strategies for operative staging to address the abdominal wall closure based on the size of the omphalocele, patient stability, and the presence of comorbid conditions or associated anomalies.
  - Recognize the need for a multidisciplinary team with experience in the repair of cloacal exstrophy and the need to transfer to a tertiary care center.
  - Counsel the family on the diagnosis of cloacal exstrophy, and discuss anatomic variations, long-term outcomes, and quality of life

❖ Intraoperative
- GS
  - Identify the safest location for the procedure (bedside vs operating room) at your institution.
  - Place a spring-loaded silo in a newborn with GS at the bedside using a silo of appropriate size.
  - Perform a sutureless umbilical closure at the bedside after identifying the instruments and supplies needed for the procedure.
  - Manage the silo using daily reduction techniques, and determine the timing for GS closure in a baby with a silo in place.
  - Safely perform a delayed GS repair using the fascial closure or sutureless umbilical closure techniques.
  - Recognize how abdominal pressure can be monitored during and after GS closure.
  - Identify the need to open the fascia more widely in certain settings.
  - Perform an assessment and initial safe management of a suspected intestinal atresia.
- Omphalocele
  - In a baby with a giant omphalocele, initiate a "paint and wait" strategy for epithelialization of the omphalocele sac and delayed ventral hernia repair.
  - In a baby with a small or medium omphalocele, safely perform a primary omphalocele repair.
  - Demonstrate a technique to perform an umbilicoplasty in an infant undergoing primary omphalocele repair.
  - In the setting of a ruptured omphalocele sac, understand the different options available for repair, including primary fascial repair, sac repair, and silo placement.
  - Demonstrate the techniques required for a complex omphalocele repair (eg, component separation).

- Cloacal exstrophy
  - Describe the goals of complete surgical repair, whether staged or primary, including securing the abdominal wall, separating the hindgut from the bladder, maintaining bowel length, and ultimately closing the bladder.

THE AMERICAN BOARD OF SURGERY

Evaluation and Management of a Patient with an Abdominal Wall Defect

- Recognize when the omphalocele cannot be closed primarily, and describe management options to include epithelialization or silo placement.
- Understand the steps for the creation of a functional hindgut ostomy, including protection of the blood supply, preservation of the hindgut, and tubularization of the cecal plate.
- Demonstrate a basic understanding of management strategies regarding conversion of the cloacal exstrophy to a classic bladder exstrophy with delayed bladder closure and osteotomies for closure of the pelvis.

❖ Postoperative

- GS
  - Formulate a nutritional plan for a baby with GS awaiting return of bowel function, including parenteral nutrition and the gradual advance of enteral nutrition.
  - In the early postoperative setting, recognize the signs of abdominal compartment syndrome, and formulate a surgical plan to reopen the abdomen.
  - Identify the natural history of umbilical hernia in a baby with GS who undergoes the sutureless umbilical closure technique, including the indications for and timing of surgical repair.
  - Identify the importance of long-term follow-up to evaluate growth, abdominal wall hernia development, and nutritional status.

- Omphalocele
  - In the early postoperative setting, recognize the signs of abdominal compartment syndrome, and formulate a surgical plan to reopen the abdomen.
  - Formulate a plan for additional staged procedures and ways to increase abdominal domain.
  - Identify the importance of long-term follow-up to evaluate growth and abdominal wall hernia development.

- Cloacal exstrophy
  - Formulate a postoperative plan to monitor fluids and electrolytes and initiate enteral feedings.
  - Recognize electrolyte imbalances that may result from a proximal colostomy (compared with a hindgut ostomy).
  - Identify the different aspects of complex wound care of a remaining omphalocele and the bladder plates if a staged approach was used.
  - Recognize and manage surgical complications from breakdown of repair of the cecal plate, omphalocele, and bladder.
  - Demonstrate understanding of long-term complications involving urinary and fecal incontinence, bowel dysmotility, permanent ostomy, and infertility.

Scope

❖ In scope

❖ Diagnoses
- Cloacal exstrophy
- GS
- Omphalocele

THE AMERICAN BOARD OF SURGERY

Evaluation and Management of a Patient with an Abdominal Wall Defect

❖ Procedures
- GS – silo followed by repair
- Omphalocele – epithelization of the sac with delayed repair
- Initial operative management of cloacal exstrophy
- Primary suture closure
- Sutureless closure
- Staged repair

❖ Special populations
- OEIS complex
- Pentalogy of Cantrell

❖ Out of scope

THE AMERICAN BOARD OF SURGERY

Evaluation & Management of a Patient with an Abdominal Wall Defect

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 assistance, performs a basic H&P and identifies the preop studies required to safely proceed with an operation in an uncomplicated patient with an abdominal wall defect. • With active assistance, formulates an initial surgical plan for an uncomplicated patient, including bedside vs. OR repair and primary vs staged/delayed repair • With active guidance, identifies and directs management for a patient who requires emergent intervention due to complicated gastroschisis, bowel ischemia, or ruptured omphalocele • Demonstrates basic knowledge of fluid losses in term neonates with abdominal wall defects • With active assistance, directs preop fluid resuscitation, temperature management, gastric decompression, patient positioning, and ventilatory strategies in an uncomplicated patient • Establishes a professional rapport with a patient and their family and communicates the basic risks and benefits of a planned procedure without diagnosis-specific details • Reviews prenatal diagnostic information without interpretation or formulation of a	• With active assistance, performs primary repair of a neonate with an uncomplicated or small abdominal wall defect • In an uncomplicated patient, requires active assistance with intraop decision-making and moving through the steps of an operation regarding the need for silo placement or widening the abdominal wall defect (GS) or delayed or staged repair, incision type, and need for patch (OM) • With active assistance, handles tissues gently and identifies normal anatomic structures, delicate/thin membranes, or tissue planes	• With active assistance, identifies the need for postop enteral or parenteral nutrition and formulates a plan • With active assistance, recognizes postop abdominal compartment syndrome and formulates a plan for management in an uncomplicated neonate • With active assistance, formulates and communicates with care teams/family on a basic plan for suspected intestinal atresia associated with gastroschisis or staged abdominal wall closure of a giant omphalocele • With active assistance in an uncomplicated neonate, recognizes postop complications (eg, wound infection, dehiscence, leak, perforation, electrolyte abnormalities, ileus, obstruction, intestinal ischemia) • With active assistance, formulates a long-term follow-up plan for evaluation of residual or recurrent hernia defects in an uncomplicated patient.

THE AMERICAN BOARD OF SURGERY

Evaluation & Management of a Patient with an Abdominal Wall Defect

Level	Nonoperative/Preoperative	Intraoperative	Postoperative
	postnatal plan and has limited participation in prenatal consultation		
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 passive assistance, performs a basic H&P and identifies the preop studies required to safely proceed with an operation in an uncomplicated patient with an abdominal wall defect. • With passive assistance, formulates an initial surgical plan for an uncomplicated patient, including bedside vs. OR repair and primary vs. staged/delayed repair. • With direct supervision, identifies and directs management for a patient who requires emergent intervention due to complicated gastroschisis, bowel ischemia, or ruptured omphalocele. • Demonstrates understanding of and describes basic aspects of fluid losses in term neonates with abdominal wall defects. • With passive assistance, directs preop fluid resuscitation, temperature management, gastric decompression, patient positioning, and ventilatory strategies in an uncomplicated patient. • Establishes a therapeutic relationship with a patient and their family and compassionately communicates the disease-specific risks and benefits of a planned procedure.	• With direct supervision, performs primary repair of a neonate with an uncomplicated or small abdominal wall defect. • In an uncomplicated patient, requires passive assistance with intraop decision-making and moving through the steps of an operation regarding the need for silo placement or widening the abdominal wall defect (GS) or delayed or staged repair, incision type, and need for patch (OM). • With passive assistance, handles tissues gently and identifies normal anatomic structures, delicate/thin membranes, or tissue planes,	• With passive assistance anticipates, formulates, and coordinates a plan for postop enteral or parenteral nutrition • With passive assistance, recognizes postop abdominal compartment syndrome and formulates a plan for management in an uncomplicated neonate • With passive assistance, formulates and communicates with care teams/family a basic plan for suspected intestinal atresia associated with gastroschisis or staged abdominal wall closure of a giant omphalocele. • With passive assistance in an uncomplicated neonate, recognizes postop complications (eg, wound infection, dehiscence, leak, perforation, electrolyte abnormalities, ileus, obstruction, intestinal ischemia). • With passive assistance, formulates a long-term follow-up plan for evaluation of residual or recurrent hernia defects in an uncomplicated patient.

THE AMERICAN BOARD OF SURGERY

Evaluation & Management of a Patient with an Abdominal Wall Defect

Level	Nonoperative/Preoperative	Intraoperative	Postoperative
	• With passive assistance, interprets prenatal diagnostic information and formulates an appropriate delivery and postnatal surgical plan with minimal participation in prenatal consultation.		
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 passive assistance, performs a detailed H&P and orders the preop studies required to safely proceed with an operation for a complex/preterm neonate with an abdominal wall defect • With passive assistance, formulates a comprehensive surgical plan for a medically complex/preterm neonate, including intraop management strategies, bedside vs OR repair, and primary vs. staged/delayed repair • With indirect supervision, identifies and directs management for a patient who requires emergent intervention due to complicated gastroschisis, bowel ischemia, or ruptured omphalocele • Demonstrates understanding of and describes comprehensive aspects of fluid losses in straight forward neonates with abdominal wall defects • With passive assistance, directs preop fluid resuscitation, temperature management, gastric decompression, patient positioning, and ventilatory strategies in a complicated patient (eg, bowel ischemia, ruptured omphalocele)	• With passive assistance, performs repair of a small abdominal wall defect in a patient presenting with gastroschisis and intestinal ischemia (eg, closing gastroschisis), small omphalocele and syndromic features, or significant comorbid conditions (complex congenital cardiac disease, pulmonary HTN, Beckwith-Wiedemann syndrome) • In a complex patient, requires passive assistance with intraoperative decision-making and moving through the steps of an operation regarding the need for silo placement, widening of the abdominal wall defect, and ostomy formation (GS) or a partial/staged omphalocele repair with attention to tissue tension, intra-abdominal pressure, and anatomic constraints (OM) • With passive assistance, handles tissues gently and identifies variations in anatomy, delicate/thin membranes, or abnormal tissue planes	• With passive assistance, anticipates, formulates, and coordinates a plan for postop enteral or parenteral nutrition in a medically or surgically complex neonate • With passive assistance, recognizes postoperative abdominal compartment syndrome and formulates a plan for management in a complex neonate • With passive assistance in a medically complex neonate, formulates a timely and comprehensive long-term surgical plan and participates in a multidisciplinary discussion with care teams/family for suspected intestinal atresia associated with gastroschisis or complex staged abdominal wall closure of a giant omphalocele • With passive assistance in a complex neonate, identifies and manages postop complications (eg, wound infection, dehiscence, leak, perforation, electrolyte abnormalities, ileus, obstruction, intestinal ischemia) • With passive assistance, formulates a long-term follow-up plan and diagnoses

THE AMERICAN BOARD OF SURGERY

Evaluation & Management of a Patient with an Abdominal Wall Defect

Level	Nonoperative/Preoperative	Intraoperative	Postoperative
	• Establishes a culturally sensitive therapeutic relationship with a socially complex family or the family of a medically complex neonate and compassionately communicates the disease-specific risks and benefits of a planned procedure and the prognosis • Independently interprets prenatal diagnostic information and formulates an appropriate delivery and postnatal surgical plan with passive participation in prenatal consultation		and manages residual or recurrent hernia defects in a complex patient
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 detailed H&P and orders the preop studies required to safely proceed with an operation any patient with an abdominal wall defect • Independently leads the formulation of a surgical plan, including intraop management strategies, bedside vs OR repair, or primary vs. staged/delayed repair in any patient with an abdominal wall defect • Independently identifies and directs management for a patient who requires emergent intervention due to complicated gastroschisis, bowel ischemia, or ruptured omphalocele • Demonstrates understanding of and describes comprehensive aspects of fluid losses in complex/preterm neonates with abdominal wall defects	• Independently performs repair of a small abdominal wall defect in a patient presenting with gastroschisis and intestinal ischemia (eg, closing gastroschisis), small omphalocele and syndromic features, or significant comorbid conditions (complex congenital cardiac disease, pulmonary HTN, Beckwith-Wiedemann syndrome) • In a complex patient, independently makes decisions intraoperatively and moves through the steps of an operation regarding silo placement, widening of the abdominal wall defect, and ostomy formation (GS) or a partial/staged giant omphalocele repair with attention to tissue tension, intra-abdominal pressure, and anatomic constraints (OM) • Adapts tissue handling to tissue type and independently identifies variations in	• Independently anticipates, formulates, and coordinates a plan for postop enteral or parenteral nutrition in a medically or surgically complex neonate • Immediately recognizes postoperative abdominal compartment syndrome and independently formulates a management plan in a complex neonate • In a medically complex neonate, independently formulates a comprehensive evidence-based and long-term surgical plan and actively participates in a multidisciplinary discussion with care teams/family for suspected intestinal atresia associated with gastroschisis or complex staged abdominal wall closure of a giant omphalocele • In a complex neonate, independently identifies and manages postop

THE AMERICAN BOARD OF SURGERY

Evaluation & Management of a Patient with an Abdominal Wall Defect

Level	Nonoperative/Preoperative	Intraoperative	Postoperative
	• Independently leads preop fluid resuscitation, temperature management, gastric decompression, and ventilatory strategies in medically complex/premature patient with an abdominal wall defect • Establishes a culturally sensitive therapeutic relationship with a socially complex family or the family of a medically complex neonate and uses shared decision-making to align patient and family values, goals, and preferences with treatment options to make a personalized care plan • Independently interprets prenatal diagnostic information and formulates an appropriate delivery and postnatal surgical plan with active participation in prenatal consultation	anatomy, delicate/thin membranes, or abnormal tissue planes	complications (eg, wound infection, dehiscence, leak, perforation, electrolyte abnormalities, ileus, obstruction, intestinal ischemia) • Independently formulates a long-term follow-up plan and diagnoses and manages residual or recurrent hernia defects in a complex patient