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Evaluation & Management of a Patient with Esophageal Atresia and Tracheoesophageal Fistula

Description of the Activity	Esophageal atresia is encountered relatively infrequently by pediatric surgeons, and its management requires meticulous clinical decision-making and operative technique. The essential functions of this activity are the establishment of effective swallowing and oral nutrition, minimization of respiratory compromise, and identification of associated congenital conditions.
Functions	❖ Nonoperative/Preoperative ➢ Perform a thorough prenatal consultation. ➢ Obtain maternal history and fetal imaging. ➢ Provide fetal risk stratification based on associated anomalies (eg, VACTERL [vertebrae, anus, cardiac anomalies, trachea, esophagus, renal anomalies, limb differences). ➢ Outline a prenatal surveillance strategy and delivery plan. ➢ Discuss the impact of polyhydramnios on the delivery plan and the potential need for intervention (eg, amnioreduction). ➢ Communicate predicted postnatal risk outcomes to the family and the multidisciplinary care team. ➢ Assist in determining when referral to a specialized center is indicated. ➢ Establish the diagnosis, and delineate the tracheoesophageal anatomy. ➢ Perform necessary preoperative investigations to inform operative timing and management options. ➢ Identify associated congenital conditions, and adapt management accordingly (eg, VACTERL). ➢ Communicate the diagnosis and treatment options to the family and other members of the health care team. ➢ Outline the timing of definitive repair (eg, delayed approach in long-gap esophageal atresia). ➢ Identify the risks and benefits of management strategies. ➢ Recognize high-risk situations (eg, very low birth weight, premature infant, complex congenital heart disease). ➢ Identify the need for and timing of additional procedures during esophageal atresia and tracheoesophageal fistula (EA/TEF) repair (eg, colostomy for anorectal malformations [ARM], duodenal atresia repair, gastrostomy). ➢ 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 ➢ Perform the procedures required to manage esophageal atresia through minimally invasive and open approaches. ➢ Confirm correct patient positioning, and ensure that the necessary equipment and materials are available ➢ Perform bronchoscopy as indicated ➢ Recognize the need for primary versus staged repair.

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	➢ Perform additional procedures when necessary (eg, colostomy for ARM, gastrostomy). ➢ Recognize and confirm all relevant anatomy. ➢ Establish clear intraoperative communication with the operating room team (anesthesia, nursing). ➢ Recognize and develop a management plan for unexpected intraoperative findings. ➢ Identify aberrant anatomy. ➢ Recognizes excessive gap or tension and institutes appropriate management to complete immediate repair vs delayed repair (eg, lengthening procedures) ➢ Recognize indications to convert from a minimally invasive to an open approach. ❖ Postoperative ➢ Communicate the postoperative plan of care to the family and other involved health care team members. ➢ Manage postoperative ventilation, timing of enteral feeds (and investigations leading to this decision), and other early complications such as hemorrhage, chylothorax, and esophageal leak. ➢ Manage postoperative esophageal strictures, including congenital esophageal stenosis. ➢ Identify long-term morbidity (chronic lung disease, gastroesophageal reflux, failure to thrive, associated congenital anomalies). ➢ Recognize the importance of a multidisciplinary approach to long-term follow-up.
Scope	❖ In scope • Diagnoses ○ Esophageal atresia ○ Tracheoesophageal fistula • Procedures ○ Bronchoscopy ○ Esophageal atresia repair ○ Esophagoscopy with or without dilation ○ TEF ligation/division ○ Thoracoscopic esophageal atresia repair • Special populations ○ Long gap, H-type ○ Recurrent TEF

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	○ Unstable patient (eg, severe respiratory distress) ○ VACTERL ❖ Out of scope • Diagnoses/procedures ○ Esophageal stricture ○ Iatrogenic tracheoesophageal fistula

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Level	Nonoperative/Preoperative	Intraoperative	Postoperative
Level 1 **Framework:** The learner demonstrates understanding of information and has basic skills What a new pediatric surgery fellow should know Entrustment: The attending will show and tell or the learner acts as first assistant.	• With active guidance, integrates prenatal and neonatal physiology and imaging studies to develop a management plan for a near/full-term infant with esophageal atresia without a major congenital condition or cardiorespiratory compromise • With active guidance recognizes the need for potential emergent evaluation of infant with esophageal atresia who has respiratory distress • With active guidance, uses language that values the neonatology and anesthesiology teams in communications regarding issues with airway and ventilatory support specific to tracheoesophageal fistula • Demonstrates knowledge of normal tracheoesophageal anatomy and development	• With direct supervision, chooses and assembles bronchoscopy equipment • Performs rigid bronchoscopy with active guidance • Serves effectively as first assistant for closure of the tracheoesophageal fistula, handling of the distal esophagus, and performance of the esophago-esophagostomy • With active guidance and normal anatomy, identifies the appropriate tissue planes during muscle-sparing thoracotomy, retropleural exposure of the tracheoesophageal fistula, dissection of the tracheoesophageal fistula, and identification of the tracheoesophageal groove • Functions as first assistant in the management of an inadvertent tracheal injury during dissection	• With active guidance, manages the postop course of a medically and surgically uncomplicated patient with esophageal atresia • With active guidance, identifies the rationale for long-term management of a patient with esophageal atresia (eg, association with gastroesophageal reflux, stricture, esophageal dysplasia)
2 **Framework:** The learner demonstrates understanding of the steps of the operation but requires direction through principles and does not	• Under direct supervision, integrates prenatal and neonatal physiology and imaging studies to develop a management plan for a near/full-term infant with esophageal atresia without a major congenital condition or cardiorespiratory compromise • Under direct supervision, recognizes the need for potential emergent evaluation of	• With indirect supervision, chooses and assembles bronchoscopy equipment • Performs rigid bronchoscopy with direct supervision, requiring assistance to identify the tracheoesophageal fistula • With indirect supervision, performs closure of the tracheoesophageal fistula, careful handling of the distal esophagus,	• With direct supervision, manages the postop course of a medically and surgically uncomplicated patient with esophageal atresia • With direct supervision, describes a general long-term management plan for a patient with esophageal atresia (eg,

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know the nuances of a basic case <u>Entrustment</u>: 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	infant with esophageal atresia who has respiratory distress, and initiates basic workup and resuscitation in a timely manner • Under direct supervision, communicates with the neonatology and anesthesia teams regarding issues with airway and ventilatory support specific to tracheoesophageal fistula • Demonstrates knowledge of abnormal tracheoesophageal anatomy and development	and esophago-esophagostomy in a stable patient • With indirect supervision and normal anatomy, identifies the appropriate tissue planes during muscle-sparing thoracotomy, retropleural exposure of the tracheoesophageal fistula, dissection of the tracheoesophageal fistula, and identification of the tracheoesophageal groove • With direct supervision manages an inadvertent tracheal injury during dissection	association with gastroesophageal reflux, stricture, esophageal dysplasia)
3 **<u>Framework:</u>** 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 <u>Entrustment</u>: The learner can perform the operation/task independently in the uncomplicated patient	• With indirect supervision, develops a plan for a patient with complicated esophageal atresia and concurrent duodenal atresia, anorectal malformation, or cardiorespiratory compromise • With indirect supervision, develops a treatment plan for a complicated case of severe respiratory distress, such as a patient with gastric distention requiring decompression, tracheomalacia, or need for emergent GE junction occlusion • With indirect supervision, communicates concerns and provides feedback to the neonatology team regarding issues with airway and ventilatory support specific to tracheoesophageal fistula	• Independently chooses and assembles bronchoscopy equipment • Performs rigid bronchoscopy and identifies the tracheoesophageal fistula with indirect supervision • With indirect supervision, performs closure of the tracheoesophageal fistula, careful handling of the distal esophagus, and esophago-esophagostomy in a complex patient (eg, cardiac anomalies, complicated anatomy) or a patient with cardiorespiratory compromise • With indirect supervision and variant anatomy, identifies appropriate tissue planes during muscle-sparing thoracotomy, retropleural exposure of	• With indirect supervision, manages the postop course of a medically complicated esophageal patient with esophageal atresia patient (eg, prematurity, congenital heart disease, anastomotic leak, early anastomotic stricture) • With indirect supervision, follows an evidence-based long-term management plan for a patient with esophageal atresia (eg, management of gastroesophageal reflux, prevention and management of stricture, screening for esophageal dysplasia)

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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, identifies surgically relevant variations in tracheoesophageal anatomy (eg, right-sided aortic arch, proximal tracheoesophageal fistula) and development and alters patient management accordingly	the tracheoesophageal fistula, dissection of the tracheoesophageal fistula, and identification of the tracheoesophageal groove • With indirect supervision manages an inadvertent tracheal injury during dissection	
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 prenatal and neonatal physiology and imaging studies to develop a management plan for a complicated infant with esophageal atresia (eg, prematurity or complex congenital heart disease) • Independently, develops a treatment plan for a complicated case of severe respiratory distress, such as a patient with gastric distention requiring decompression, tracheomalacia, or need for emergent GE junction occlusion • Independently coordinates recommendations from the neonatology or anesthesiology team regarding issues with airway and ventilatory support specific to tracheoesophageal fistula in a crisis situation • Independently identifies surgically relevant variations in tracheoesophageal anatomy and development (eg, right-sided aortic arch, proximal tracheoesophageal fistula) and alters patient management accordingly	• Independently chooses and assembles bronchoscopy equipment, including additional equipment needed for a complicated case (eg, Fogarty, concomitant esophagoscopy) • Independently moves forward with rigid bronchoscopy and with passive supervision refines the approach in a complicated situation (eg, hypoxemia, excessive secretions) and identifies abnormal anatomy (eg, congenital subglottic stenosis, laryngotracheoesophageal cleft) • Independently performs closure of the tracheoesophageal fistula, handling of the distal esophagus, and esophago-esophagostomy in a complex patient • Independently visualizes tissue planes and performs muscle-sparing thoracotomy, retropleural exposure of the tracheoesophageal fistula, dissection of the tracheoesophageal fistula, and identification of the tracheoesophageal groove	• Independently manages the postop course of a medically complicated esophageal atresia patient (eg, prematurity, congenital heart disease, anastomotic leak, early anastomotic stricture) • Integrates patient- and family-specific factors into an evidence-based long-term management plan for a patient with esophageal atresia (eg, management of gastroesophageal reflux, prevention and management of stricture, screening for esophageal dysplasia)

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		• With independently manages an inadvertent tracheal injury during dissection	