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Evaluation and Management of a Patient with an Anorectal Malformation

Description of the Activity	Pediatric surgeons must be able to evaluate and manage pediatric patients with an anorectal malformation (ARM), from prenatal counseling to reconstructive procedures, to adolescence, in a variety of practice settings. The key features of ARM include: • A spectrum of anomalies with multiple subtypes ranging from mild to severe, depending on the specific nature of the malformation ○ Some cases may involve only minor abnormalities that can be corrected with relatively simple surgical procedures, while others may require more complex surgical interventions. • Abnormalities in the genitourinary tract, such as the urinary and reproductive organs • Associated conditions, such as VACTERL (vertebral defects, anal atresia, cardiac defects, tracheoesophageal fistula, renal anomalies, and limb abnormalities The prognosis and long-term outcomes for pediatric patients with ARM depend on the severity of the malformation, the success of surgical interventions, and the presence of any associated conditions. Early detection and intervention are crucial for optimizing outcomes and improving the quality of life for affected individuals.

Functions	❖ Nonoperative/Preoperative
	
	• Identify common findings on prenatal ultrasound in a patient with cloaca, including abdominal/pelvic cystic masses, hydronephrosis, hydroureteronephrosis, oligohydramnios, ascites, and distended or obstructed bowel.
	  ○ These findings should prompt additional evaluation with the maternal-fetal medicine team and may prompt magnetic resonance imaging (MRI).
	  ○ If MRI findings suggest cloaca or ARM, refer the patient to a center for counseling, and consider delivery at an appropriate facility.
	• Counsel parents and families on delivery expectations and the anticipated postnatal course.
	• Obtain a thorough history and physical exam, identifying prenatal risk factors. Differentiate between common and uncommon ARM variants on exam.
	• Communicate the clinical workup, evaluation, and expectations to the parents and other health care providers.
	• Perform a VACTERL workup, and identify important comorbid conditions.
	• Identify a unique ARM problem (eg, hydrocolpos in the setting of cloaca and potential complications).
	• Recognize factors that portend outcome, such as tethered cord, high versus low defect, and sacral ratio.
	• Perform initial nonoperative and operative management
	• Identify the role of early versus delayed repair in patients with perineal and vestibular fistulas.
	• Identify the indications and outline the technical aspects for a diverting colostomy in the neonatal period.
	• Evaluate a patient with cloaca, and identify initial operative management strategies.
	• Evaluate and manage a patient with hydrocolpos.
	• Perform panendoscopy and a three-dimensional cloacagram.
	• Perform and interpret a distal high-pressure colostogram, recognizing its impact on surgical management.
	• 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.

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- 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
- Describe the necessary steps for operative repair.
- Ensure the patient is positioned correctly and the operating room contains the necessary equipment and materials (eg, Foley catheter, antibiotics).
- Identify the sphincter complex with electrostimulation.
- Describe the key steps of a posterior sagittal anorectoplasty (PSARP) in male and female patients.
- Manage any urethral anomalies (e.g. rectourethral fistula).
- Identify the key steps of and indications for laparoscopic repair of ARM.
- For a patient with cloaca:
  - Identify the initial need for colostomy, and describe its technical aspects.
  - Describe the operative management for likely scenarios.
  - For a patient with type 1 cloaca:
    - Common channel < 1 cm: urethra is left untouched, introitoplasty, and PSARP
    - Common channel < 3 cm and urethra > 1.5 cm: total urogenital mobilization (TUM) and PSARP
    - Common channel > 3 cm or urethra < 1.5 cm: urogenital (UG) separation with the common channel as the urethra and PSARP
- Recognize the risk to the ureter during dissection.
- Identify options for vaginal replacement.

❖ Postoperative
- Recognize the rationale for the debate regarding time to postoperative feeds in a nondiverted patient and the need for postoperative anal dilations.
- Perform postoperative management for a patient with a Foley catheter who has undergone ARM repair.
- Manage postoperative complications (eg, surgical site infection [SSI] in a nondiverted PSARP).
- Identify indications for urologic and gynecologic follow-up and evaluation.
- Identify possible long-term outcomes.
- Evaluate a patient who is “not doing well.”
- Discuss problems with the prior diagnosis, decision-making, studies, or operative issues.
- Recognize and address common post-operative problems:
  - Constipation
  - Fecal incontinence
  - Fistulae
  - Mislocated anus
  - Obstructed menstruation
  - Persistent UG sinus

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	▪ Retained remnant of original fistula ▪ Rectal prolapse ▪ Stricture of the rectum/neoanus ▪ Urethra atresia/stricture/injury ▪ Urinary tract infection ▪ Vaginal atresia • Determine the role of bowel management.
Scope	❖ In scope • Diagnoses ○ Male and female: ▪ Low and high ARMs ○ Gender nonspecific: ▪ Rectoperineal ▪ Without fistula ○ Male: ▪ Bladder neck ▪ Rectourethral (bulbar/prostatic) ○ Female: ▪ Cloaca (short/long) ▪ Cloacal exstrophy ▪ Rectovestibular • Procedures ○ Anoplasty ○ Antegrade continence enema ○ Cloacal repair ○ Diverting colostomy ○ Laparoscopic-assisted anorectal pull-through ○ PSARP/anterior sagittal anorectoplasty • Special populations ○ Currarino syndrome ○ VACTERL (complex decision-making)

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❖ Out of scope

- Diagnoses/procedures
  - Bladder exstrophy
  - Disorder of sexual development
  - Fissures
    Fistula-in-ano
  - Hemorrhoids
  - Pilonidal disease
  - Rectal prolapse

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Level	Nonoperative/Preoperative	Intraoperative	Postoperative
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 assistance, elicits a focused H&P that includes a prenatal history and correct identification of the anatomy • With active assistance, initiates an appropriate workup/evaluation, including VACTERL anomalies, and discusses the role of diversion or dilations • With active guidance, identifies the appropriate surgical options for a common ARM variant (eg, PSARP, laparoscopy, primary PSARP vs delayed PSARP) • Respectfully communicates basic facts about the condition to the family and interprofessional team	• With active guidance, assists in the management of the periop environment, including room setup, equipment check, time-out, patient positioning, and debrief • With active guidance, progresses through the creation of a stoma, requiring assistance on technical nuances (eg, location, distal decompression) • With active guidance, progresses through major steps of a straightforward PSARP for a perineal or vestibular fistula and critical aspects of the operation, including use of a stimulator to identify the sphincter complex; requires active assistance to stay in the appropriate plane and recognize major structures at risk (male urethra, posterior vagina, fistula if present) • With active assistance, manages, anticipates, and prevents intraop complications for a common procedure (eg, enterotomy, bleeding)	• Recognizes and manages a general postop problem (eg, SSI, skin dehiscence) but displays limited understanding of an ARM-specific complication (eg, anal stenosis, mucosal prolapse) • Displays limited knowledge of long-term treatment/outcomes (eg, bowel management) • With active assistance, manages a patient's postop course following a PSARP for a perineal fistula • Communicates basic aspects of the operative procedure and ongoing management plan to the family and interprofessional team
2 **Framework:** The learner demonstrates understanding of the steps of the operation but requires direction through principles and does not	• With direct supervision, elicits a focused and complete H&P with identification of a common anatomic variant and articulates a management strategy • With indirect supervision, integrates information with patient-specific factors to design a succinct diagnostic workup	• With direct supervision, assists in the management of the periop environment, including room setup, equipment check, time-out, patient positioning, and debrief • With passive guidance, progresses through the creation of a stoma	• Recognizes an ARM-specific complication (eg, anal stenosis, mucosal prolapse) but requires direct supervision to institute a treatment plan • Demonstrates understanding of long-term outcomes and the role of bowel

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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	and management plan for a medically uncomplicated surgical patient • With direct supervision, identifies the appropriate surgical options for a common ARM variant (eg, PSARP, laparoscopy, primary PSARP vs delayed PSARP) • Establishes a therapeutic relationship with the family of a uncomplicated patient and communicates information clearly to the care team	• With direct supervision, progresses through major steps of a straightforward PSARP for a perineal or vestibular fistula and critical aspects of the operation, including use of a stimulator to identify the sphincter complex; requires passive assistance to maintain dissection in the optimal tissue plane and may inconsistently demonstrate careful handling of tissue • With passive assistance, manages, anticipates, and prevents intraop complications for a common procedure (eg, enterotomy, bleeding)	management but needs direct supervision in implementing long-term therapy and follow-up • With indirect supervision, manages the postop course of a patient with a common ARM but requires direct supervision for a complex or uncommon ARM • Communicates a cohesive postop plan to the family and interprofessional team for a common ARM variant
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	• With indirect supervision, elicits a focused and complete H&P with identification of an uncommon anatomic variant and articulates a management strategy • With indirect supervision, initiates an appropriate workup/evaluation for a medically complicated patient (eg, congenital heart disease, multiple anomalies) • With indirect supervision, identifies the appropriate surgical options for an uncommon ARM variant, including management of a clinically important comorbid condition (eg, hydrocolpos)	• With indirect supervision, manages the periop environment, including room setup, equipment check, time-out, patient positioning, and debrief • With indirect supervision, progresses through the creation of a stoma • With indirect supervision, progresses through major steps of PSARP, including an advanced case such as a rectourinary fistula in males, possibly including open or laparoscopic approaches; requires direct supervision to maintain dissection in the optimal tissue plane; visualizes tissue planes and identifies and dissects relevant normal anatomy and consistently demonstrates careful handling of tissues	• With indirect supervision, recognizes, but does not anticipate, an ARM-specific complication (eg, anal stenosis, mucosal prolapse) and institutes a proper treatment plan • Articulates the need for long-term follow-up and with assistance implements a bowel management program with indirect supervision • With indirect supervision, manages the postop course of a complicated neonatal or pediatric surgical patient

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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	• Establishes a therapeutic relationship with the family of a complicated patient and uses active listening skills to adapt to care team needs	• With indirect supervision, recognizes and manages findings such as an unexpected fistula or iatrogenic injury to the vagina or urethra and implements modifications to the operative plan in real time	• Communicates a cohesive postop plan to the family and interprofessional team for an uncommon ARM variant
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 assesses and performs a focused and complete H&P, interprets diagnostic images (e.g. colostogram), reviews reports, and initiates an individualized management plan across the spectrum of ARMs, addressing clinically important comorbid conditions (eg, hydrocolpos) • Independently recognizes limitations (if applicable) and engages additional resources required for the care of a complex patient (eg, additional consultants, engagement with a colorectal center) • Independently identifies the appropriate surgical options for an uncommon ARM variant, including management of a clinically important comorbid condition (eg, hydrocolpos) • Independently conducts a family/care team meeting to define the goals of care	• Independently manages the periop environment, including room setup, equipment check, time-out, patient positioning, and debrief • Independently performs and properly locates a diverting ostomy • Independently progresses through major steps of PSARP, including an advanced case such as a rectourinary fistula in males and short common channel cloaca in females, possibly including open or laparoscopic approaches; visualizes tissue planes and identifies and dissects relevant abnormal anatomy • Independently recognizes and manages findings such as an unexpected fistula or iatrogenic injury to the vagina or urethra and implements modifications to the operative plan in real time	• Independently anticipates and implements a strategy for management of a postop complication, such as dehiscence of perineal body, mislocated anus, fistulae, ROOF, rectal prolapse, or urethral injury or stricture • Independently initiates a postop plan that may include a bowel management program or multidisciplinary involvement to assist with comorbid conditions • Independently manages the postop course of a complicated neonatal or pediatric surgical patient • Independently customizes emotionally difficult news such as changes to the operative plan, adverse outcomes, expectations, or additional procedures to the family in a culturally caring manner

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	for a complicated pediatric surgical patient, including informed consent		