Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Pharyngitis (primary)		Cervical adenopathy Mucosal ulceration	
GU (mucous membranes)	Cystitis (primary) Genital ulcers (recurrent)	Dysuria	Vaginal or urethral discharge Ulcerated lesions of penis or labia or cervix	Viral culture (gold standard) Tzanck smear; direct immunofluorescent assay Biopsy; intranuclear inclusion bodies
LYMPH		Lymphadenopathy	Tender inguinal nodes	
DERM	Herpetic whitlow (recurrent)	Painful vesicular or papular lesion	Pain	Tzanck smear
CNS	Aseptic meningitis (primary)	Headache	Cauda equina syndrome	
RECTAL	Herpes proctitis (primary)	Constipation Tenesmus Discharge		Proctosigmoidoscopy

Key References: Armitage KB, Salata RA: Sexually transmitted diseases. In Andreoli TE, Carpenter C, Griggs RC, editors: *Andreoli and Carpenter's Cecil essentials of medicine,* ed 7, Philadelphia, PA, 2007, Saunders, pp 980–988; Augenbraun M, Feldman J, Chirgwin K, et al.: Increased genital shedding of HSV-2 in HIV-seropositive women, *Ann Intern Med* 123(11):845–847, 1995.

Perioperative Implications

Preoperative Preparation
- Universal precautions

Monitoring
- Routine

Regional Anesthesia
- Needle placement in infected area contraindicated secondary to risk of viremia and local extension into deep tissues
- Preferred in pregnant women with recurrent infection, no systemic symptoms, and no infection in area of block placement

Postoperative Period
- Universal precautions

Anticipated Problems/Concerns
- Difficulty identifying asymptomatic carriers of HSV-2 with viral shedding
- No effective prophylaxis for newborns

Hirschsprung Disease

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Risk
- Incidence of 1:5000 live births; varies among different ethnic groups.
- Male to female ratio is 4:1, although bias is lost in longer segment disease.
- Occurs as an isolated phenotype but may be associated with congenital abnormalities and associated syndromes.
- Up to 30% of affected individuals have at least one coexisting congenital anomaly, which may include congenital heart defects, gastrointestinal malformations, central nervous system, genitourinary, and craniofacial abnormalities, and spina bifida.
- Between 2-15% of affected individuals have trisomy 21. Other associated syndromes include Waardenburg syndrome type IV, congenital central hypoventilation syndrome (Ondine's Curse), multiple endocrine neoplasia type 2, and neurofibromatosis.

Perioperative Risks
- Intestinal obstruction.
- HAEC, characterized by explosive foul-smelling diarrhea, abdominal distension, and fever, may progress to potentially fatal toxic megacolon.
- Septic shock.
- Hypovolemia.
- Lyte abnormalities.

Worry About
- Intestinal obstruction increases the risk of regurgitation and pulmonary aspiration.
- Vomiting and possible diarrhea leads to hypovolemia and lyte abnormalities, necessitating adequate resuscitation.
- Pts presenting with HAEC may have septicemia requiring preop antibiotic administration.

Overview
- HSCR is a multigenic disorder with variable penetrance. In over 80%, aganglionosis is restricted to the rectosigmoid colon (short segment HSCR), but may affect significant lengths of colon and even extend into the small intestine (long segment HSCR) or rarely in 3–8% affects the entire small and large intestines (total intestinal aganglionosis).
- Most often, pts are diagnosed in the neonatal period with distended abdomen, delayed passage of meconium (>24–48 h), and vomiting.
- Pts diagnosed later in childhood present with chronic constipation and failure to thrive.
- Diagnosis can be made with plain radiography (with marked gaseous distension of colon and an undilated rectum), contrast enema (which defines the transition zone between dilated normal bowel and narrow aganglionic bowel), and anorectal manometry.
- Gold standard for diagnosis is a rectal biopsy (submucosal suction or full thickness), demonstrating absence of ganglion cells and presence of acetylcholinesterase-positive hypertrophic nerve fibers.

Etiology
- Initial symptoms are caused by failure of neural crest cells to caudally migrate and colonize variable lengths of the intestinal tract.
- Complete absence of enteric neuronal ganglion cells in the affected bowel results in tonic contraction, leading to obstructive symptoms.
- Several key genes regulating neural crest cell development, including *RET*, *GDNF*, and *EDNRB*, are associated with HSCR but only account for about 50% of known cases.
- Combinations of genetic mutations and modifiers likely contribute to etiology and pathogenesis.

Usual Treatment
- If neonate presents with enterocolitis, aggressive resuscitation, rectal irrigation, and antibiotics are utilized for initial management.
- Surgery, the only definitive treatment for HSCR, aims to remove aganglionic bowel and anastomose normal bowel to the anus while preserving sphincter function.
- Transition zone identified by intraop frozen sections sent to pathology may determine operative time.
- Traditionally, operative repair was performed in two or three stages. First stage required a diverting ostomy, second stage (usually at 3 mo-1 y of age) involved resection of aganglionic bowel and coloanal anastomosis, and third stage entailed closure of preexisting stoma.
- Classical operations (Swenson, Soave, Duhamel) are now reduced to one or two stages. Standard approach in otherwise healthy, nondistended neonatally diagnosed HSCR is a one-stage repair.
- TERPT or LATEP is associated with faster recovery, shorter hospital stay, and fewer postop complications.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
RESP	Congenital hypoventilation ("Ondine's curse")	Apnea		
CV	Hypovolemia, septic shock, 2-5% cardiac anomalies (tetralogy of Fallot)	IV replacement Extent of vomiting Cyanosis	Mucous membranes Vital signs/UO Murmur, cyanosis Capillary refill	BUN, Cr BUN/Cr ratio ECHO
GI	Intestinal obstruction	Presence of meconium Constipation Diarrhea Vomiting	No feces in rectum, Abdominal distention Malnutrition	Lyte panel Abdominal films Barium enema

Key References: Butler Tjaden NE, Trainor PA: The developmental etiology and pathogenesis of Hirschsprung disease, *Transl Res* 162(1):1–15, 2013; McKeown SJ, Stamp L, Hao MM, et al.: Hirschsprung disease: a developmental disorder of the enteric nervous system, *Wiley Interdiscip Rev Dev Biol* 2(1):113–129, 2013.

Perioperative Implications

Preoperative Preparation

- Consider associated congenital anomalies or syndromes and the possible need for further cardiac evaluation and genetic testing.
- Thoroughly assess volume status. Assess for bowel preparation, diarrhea, and vomiting and ensure adequate preop fluid resuscitation.
- Review preop labs to assess for lyte abnormalities.
- Consider cardiac evaluation with associated cardiac anomalies.

Monitoring

- Standard ASA monitors
- Urinary cath

Airway

- Consider associated syndromes affecting airway anatomy.

Induction

- Rapid sequence induction necessary in the presence of bowel obstruction to avoid pulmonary aspiration.
- In the setting of hypovolemia or sepsis (HAEC), IV and volatile anesthetics may be poorly tolerated.

Maintenance

- Use neuromuscular blocking drugs for maintenance of muscle relaxation.
- Maintenance IV fluids with balanced, isotonic solution.
- Consider checking intraop blood glucose level.
- Monitor urine output.
- Avoid nitrous oxide.
- Maintain normothermia with warming devices (full access warming blankets and radiant warmers), as radiant heat loss may be excessive. Keep forced warm air blankets dry. They cool the pt if they become wet.
- Carefully position pt; use added care with lithotomy position.

Extubation

- Reverse neuromuscular blockade.
- Routinely extubate when pt is awake and meets extubation criteria.

Postoperative Period

- Consider regional technique with epidural/caudal anesthesia for postop pain management (which may need to be performed postop if lower body antibacterial preparation performed).
- Postop apnea in newborns more likely following narcotic administration.

Anticipated Problems/Concerns

- Early postop complications include prolonged ileus, anastomotic leak, and wound infection/dehiscence.
- Late complications include anastomotic strictures, constipation, fecal incontinence, bowel obstruction, and enterocolitis.
- Postop HAEC, with an incidence between 5-42%, is a major cause of increased morbidity and mortality after definitive pull-through procedure. This is hypothesized to involve intestinal stasis and immature mucosal immunity, allowing for proliferation and mucosal invasion by luminal pathogens.
- Mild obstructive symptoms are treated with dietary changes, laxatives, enemas, or repeated botulinum toxin injections. Myectomy procedure may be required.
- For residual aganglionosis, strictures, or dysfunctional proximal bowel, repeat pull-through procedure can be done, although this is challenging due to scarring.
- In individuals with extensive intestinal aganglionosis and irreversible intestinal failure, intestinal transplantation may be considered.

Histiocytosis

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Risk

- LCH is the most commonly known form.
- Incidence: 1:250,000 in children, with about a third of this incidence in adults.
- Seen in all ages, but peak incidence is at 0–3 y of age.
- Male:female ratio: 1.5:1.
- Sporadic development with no established genetic predisposition.

Perioperative Risks

- Dependent on organ systems involved and extent of dysfunction

Worry About

- Specific organ dysfunction caused by infiltration with histiocytes, including liver, lungs, hematopoietic system, pituitary, spleen, and bone
- Can involve single or multiple sites and organs
- Treated with steroids and chemotherapy, which may cause adrenal insufficiency and result in the pt requiring stress steroids in the periop period
- Central diabetes insipidus due to posterior pituitary involvement
- Cervical instability if lesions present in cervical vertebrae
- Severe pulm dysfunction possible; pulm Htn without overt right heart failure

Overview

- A broad group of disorders involving infiltration of affected organs with monocytes, macrophages, and dendritic cells.
- The most commonly discussed disorder is LCH, previously known separately as eosinophilic granuloma, Hand-Schüller-Christian disease, and Letterer-Siwe disease.
- Severity of clinical symptoms varies markedly and can involve primarily skin and/or bone or liver, lung, or brain.
- Can be limited or progressive and fatal. Younger children with multiple or severe organ involvement of "risk organs" (liver, lungs, spleen, hematopoietic system) have a high mortality.
- Usual clinical presentation is in the first decade of life.
- Pathophysiology is unclear and treatment is nonspecific.

Etiology

- Unknown; suggested factors include immune dysfunction, viral infections, neoplastic processes, and genetic predisposition.
- Isolated pulm LCH is strongly associated with cigarette smoking.

Usual Treatment

- 10–20% spontaneous regression rate, almost exclusively in pts with single system disease.
- Chemotherapy with steroids for multisystem disease with local or constitutional symptoms (vinblastine, etoposide, mercaptopurine, doxorubicin, cyclophosphamide, methotrexate, others).
- Surgery is required for biopsy and Dx, isolated bone lesions, and occasionally splenectomy.
- Orthotopic liver or lung transplantation has been performed for end-stage disease.
- Radiation therapy (bone lesions, pituitary disease).
- Bone marrow or stem cell transplant.