Assessment Points

System	Effect	Assessment by Hx	PE	Test
RESP	Pneumonitis	Aspiration of oral secretions; previous HSV esophagitis	Bilateral crackles	CXR (bilateral interstitial infiltrates)
GI	Esophagitis	Odynophagia, dysphagia, substernal pain	Multiple shallow mucosal ulcers	
GU	Cystitis			
CNS	Encephalitis, meningitis	Headache, confusion, lethargy	Anosmia, memory loss, expressive aphasia, focal seizures	Brain biopsy
DERM	Cutaneous ulcers	Recurrent painful skin or mucosal ulcers	Multiple vesicular lesions on an erythematous base with subsequent ulceration	
	Stevens-Johnson syndrome	Extensive painful skin lesions	Deep bullous erosive lesions	

Key References: Chayavichitsilp P, Buckwalter JV, Krakowski AC, et al.: Herpes simplex, *Pediatr Rev* 30(4):119–129, 2009; Davies PW, Vallejo MC, Shannon KT, et al.: Oral herpes simplex reactivation after intrathecal morphine: a prospective randomized trial in an obstetric population, *Anesth Analg* 100(5):1472–1476, 2005.

Perioperative Implications

Preoperative Preparation

- Cover exposed herpetic lesions.
- Strict adherence to universal precautions.

Monitoring

- Avoid disturbing active lesions.

Regional Anesthesia

- Needle should not be inserted through lesion. There is a theoretical risk of spreading herpes from one infected ganglion to another, but regional anesthesia is not contraindicated.
- Neuraxial morphine remains a common practice, as rare occurrence of vertical transmission from mother to neonate does not support withholding this technique.

Postoperative Period

- Thoroughly disinfect any surface area that might have been in contact with oral secretions or herpetic lesions. Most disinfectants are effective, including chlorine and alcohol.

Anticipated Problems/Concerns

- No effective preexposure or postexposure prophylaxis.
- Acyclovir may reduce effectiveness of phenytoin.
- C-section should be offered for pregnant women with active HSV.
- Vaginal delivery is acceptable for women in remission; acyclovir is often used.

Herpes, Type II

Jonathan G. Ma | Alan David Kaye | Amit Prabhakar

Risk

- Incidence within USA of HSV-2 is estimated at 40–60 million (20% of sexually active adults).
- Approximately 536 million people (16% of population) infected worldwide, most unaware of the disease.
- Highest prevalence in women, African Americans, and lower socioeconomic groups.
- Frequency and severity of infection increases in immunocompromised pts, including HSV encephalitis.
- Incidence of neonatal HSV infection is estimated at 1:2000-5000 deliveries.

Perioperative Risks

- Vertical transmission from infected mother to fetus during vaginal birth
- Intrauterine fetal infection after rupture of membranes

Worry About

- Transmission of infection to health care personnel resulting in herpetic whitlow via inoculation of virus into digits is very well described and completely preventable with universal precautions (e.g., gloves at all times).
- Neonatal herpetic infection during vaginal births.
- Viremia secondary to needle placement within infected area during regional anesthesia with possible extension of infection to adjacent areas.
- Secondary bacterial or fungal infection of herpetic lesions.

Overview

- Primarily caused by infections below the waist transmitted by sexual contact.
- Maternal primary HSV-2 infection is associated with spontaneous abortion.
- Newborns can be infected with HSV-2 during vaginal delivery from the mother's genital infection (high neonatal mortality).
- Primary genital HSV-2 infection has the highest incidence of systemic symptoms (malaise, fever, headache, myalgias).
- Latent infection remains dormant in sensory ganglia, innervating the infected area until reactivation.
- Recurrent infection involves clusters of genital sores (papules and vesicles) on outer surface of genitals, usually appearing 4–7 d post HSV exposure.
- No increased risk of reactivation of HSV-2 is associated with neuraxial anesthesia.
- Chronic recurrent HSV-2 infection is associated with development of cervical and vulvar cancer.
- Reactivation is known to occur with exposure to UV light, immunosuppression, trauma, and fever.
- Dx by viral culture (gold standard) is the most sensitive and specific (rapid Dx by Tzanck smear).
- Genital herpes increases the risk of transmission and acquisition of HIV-1 infection threefold to fourfold.

Etiology

- Double-stranded DNA virus in the family of Herpesviridae.
- Acquired via genital infection primarily by sexual transmission of HSV-2.
- Immunosuppression and increased number of sexual partners are risk factors for acquisition.
- Diagnosed by multinucleated giant epithelial cells (polykaryocytes) with intranuclear (Cowdry type A) inclusion bodies on Giemsa stain smears (Tzanck preparation) taken from vesicle or tissue biopsy.

Usual Treatment

- Administer IV acyclovir for neonatal HSV-2 infection.
- Oral acyclovir and topical cream shorten duration of lesions for recurrent infections.
- Most recommend that full-term parturients with visible genital lesions (especially primary infection) undergo cesarean delivery to decrease incidence of neonatal HSV infection. Neonates exposed to asymptomatic shedding of HSV during parturition (fourfold increase in HIV seropositive women) may also rarely acquire neonatal HSV.
- Active genital herpes lesions are indications for cesarean delivery for prevention of neonatal herpes infection. This significantly reduces risk of transmission. Use of third trimester oral suppression for outbreak prophylaxis is effective at reducing risk of needing cesarean delivery.

Novel Therapies

- Pericoital application of tenofovir gel showed reduction in HSV-2 acquisition, decreased shedding, decreased lesion rate, and decreased quantity of viral shedding.
- Administer imiquimod for acyclovir-resistant HSV-2.
- The combination of imiquimod immunomodulator, imiquimod, and acyclovir appears to provide effective therapy for acute genital HSV-2 infection, even when begun after lesion development.

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

Franklyn P. Cladis | Annie Lynn Penaco

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.