

Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Macroglossia	Hx of difficult mask ventilation and intubation	Determine extent by physical inspection and oral palpation, previous anesthesia Hx	No testing
CV	VSD, ASD, TOF, valvular disease, hypoplastic LV, cardiac tumor and cardiomegaly (most common) possible	SOB, DOE	Cardiac exam for murmurs	ECHO CXR
ENDO	Hypoglycemia Hypothyroidism	Shaking, lethargy		Glucose Thyroid function tests
RENAL	Renal medullary dysplasia Nephrolithiasis	Hx of renal tumors/previous resections, chronic UTIs	Palpate for masses, Flank pain	US BUN/Cr

Key References: Weksberg R, Shuman C, Beckwith JB: Beckwith–Wiedemann syndrome, *Eur J Hum Genet* 18(1):8–14, 2010; Eaton J, Atilas R, Tuchman JB: GlideScope for management of the difficult airway in a child with Beckwith–Wiedemann syndrome, *Paediatr Anaesth* 19(7):696–698, 2009.

Perioperative Implications

Preoperative Preparation

- Coordinated care with endocrinology and an ENT specialist to assist in the management of hypoglycemia and difficult airway.
- Discussion with ENT for planned tracheostomy if significant airway edema and swelling is anticipated following glossal resection.
- Review of lab results (hypothyroidism, polycythemia, hypocalcemia, and hyperlipidemia have been reported in pts with BWS in addition to hypoglycemia).
- Review cardiac workup if available.
- Pretreatment with antisialagogue (glycopyrrolate or atropine) if intubation is planned.

Monitoring

- Standard monitoring appropriate for surgical procedure
- Frequent glucose checks

Airway

- Assume difficult mask ventilation due to macroglossia.
- Nasal intubation may be more easily performed than oral intubation in pts with significant macroglossia. Pretreat with a nasal decongestant and dilate with nasal trumpets if nasal intubation is considered.
- Assistance with glossal manipulation if direct laryngoscopy is performed.
- Backup airway devices (e.g., fiberoptic, glidescope, LMA) and surgical support (ENT) if conventional laryngoscopy fails.
- Age-appropriate ETT.

Induction

- Inhaled induction with sevoflurane versus awake intubation with sedation/topicalization.
- Clinicians should be aware that administration of IV anesthetics and muscle relaxants may cause pt's tongue to fall backward, causing acute airway obstruction.

Postoperative Period

- After meeting strict extubation criteria, pts should be monitored in ICU or recovery area with immediate backup for management of airway issues and hypoglycemia.

Anticipated Problems/Concerns

- Difficult airway
- Hypoglycemia

Behçet Disease

Anurag Gupta

Risk

- Affects age group between 20–40 y
- Nations along Silk Route have higher incidence
- Males and females are equally affected

Perioperative Risks

- Increase in IOP during intubation in pts with uveitis complicated by glaucoma
- Pulmonary embolism
- Difficult airway due to oral inflammation

Worry About

- Difficult airway
- Hyperreactive skin
- Pulmonary aneurysm
- Intracranial Htn
- Concurrent anti-inflammatory medications

Overview

- Multisystem inflammatory disorder of unknown etiology characterized by relapsing episodes of oral aphthous ulcers, genital ulcers, other skin lesions, and ocular lesions.

- For diagnosis of BD, an international study group proposed the presence of oral aphthous ulcers and any two other manifestation among the following:
 - Recurrent genital ulceration.
 - Skin lesion.
 - Papulopustular lesions.
 - Ocular involvement.
- Positive pathergy test: Hyperreactivity of skin leading to sterile pustule and erythematous papule formation after intracutaneous injection or needle prick.
- Ocular and vascular involvement increases morbidity.
- Major vessel disease and neurologic involvement are the major cause of death.
- Newer drugs have shown good improvement in resistant cases of BD, but further studies are needed to reinstate their efficacy.
- Extracutaneous ulcers, which heal by scarring, may be found in children.
- Erythematous nodosum-like lesions may occur mostly in females; the lesions are more erythematous and edematous, and they heal within a week, leaving hyperpigmentation after healing.

Etiology

- Although exact etiology unknown, BD is found to be mostly associated with HLA-B51.
- Other genes implicated are HLA-26, PSOR1C1, HLA-Cw1602, GIMAP, UBAC2, IL-10, and IL-23.

Treatment

- Mucocutaneous BD: Thalidomide, dapsone, TNF- α inhibitor (Etanercept), IFN- α , and colchicine
- Ocular involvement: Azathioprine, cyclosporine, IFN- α , and methotrexate
- Vascular involvement: Azathioprine, cyclophosphamide, and cyclosporine
- Joint involvement: Colchicine and NSAIDs
- GIT: Azathioprine
- CNS: Anticoagulants, infliximab, IFN- α , and adalimumab
- Corticosteroids: Severe, life-threatening disease in ocular, vascular, GI, and neuro BD (as advised by an expert committee report)

Assessment Points				
System	Effect	Assessment by Hx	PE	Test
HEENT	Recurrent oral aphthous ulcers Airway edema Relapsing uveitis Glaucoma Cataract Loss of vision	Frequent attacks >3 times/y Ulcers may be single/ multiple After local trauma/dental extraction Presents 2-3 y after oral lesions Frequent attacks leads to complications	Minor ulcers: <1 cm; heal without scarring in 4-14 days Major ulcers: >1 cm; heal with scarring Site involves gingival, buccal, labial mucosa, soft and hard palate Airway edema Erythematous border around ulcers Blurred vision Lacrimation Floaters Periorbital global pain Hyperemia	PCR CBC Tzanck smear HPE Vit B12 Iron Folate Intraocular fluid: • Culture • PCR • Immuno-histochemical study
RESP	Pulmonary artery aneurysm Vena cava thrombosis Pleural effusion Pulmonary infarct Pulmonary fibrosis	Previous history of oral and ocular involvement Hemoptysis Chest pain	Superior vena cave syndrome Stony dullness on percussion ↓ breath sounds	CXR CT angiography
CV	Cardiac involvement is uncommon Myocarditis Valvular lesion Pericarditis Ventricular aneurysm Vasculitis is common Venous > arterial Superficial thrombophlebitis Venous thrombosis	History of oral/ocular lesions Tenderness over the peripheral vein Calf tenderness	Dyspnea Fatigue Pulmonary edema Chest pain	ECG 2D-ECHO Doppler study of deep veins
GI	Ileitis Colitis Intestinal perforation Acute abdomen	Positive previous history Pain in abdomen Diarrhea Dyspepsia Vomiting Dysphagia Retrosternal chest pain Hematemesis	Abdominal tenderness Rigidity and guarding	GI endoscopy • Round and oval ulcers • No granulations
GU	Genital ulceration	History of aphthous ulcers Involvement of scrotum, glans or shaft in males Involvement of labia in females	Larger and deeper lesion with irregular border Heals with scar	
CNS	Meningoencephalitis Stroke in young pt Movement disorder Dural sinus thrombosis Arterial vasculitis Aseptic meningitis Intracranial and extracranial aneurysm	Headache Seizure Brain stem syndrome Ataxia Aphasia Pseudobulbar palsy Cranial nerve palsy Pyramidal tract sign		CSF examination • Increased protein • Increased cell count • Normal glucoseMRI Diffusion weighted image: Increased diffusion coefficient in BD HPE: Perivascular lymphatic infiltration with area of necrosis

Key References: Saleh Z, Arayssi T: Update on the therapy of Behçet disease, *Theor Adv Chronic Dis* 5(3):112–134, 2014; Kokturk A: Clinical and pathological manifestations with differential diagnosis in Behçet's disease, *Pathol Res Int* 2012:690390, 2012.

Perioperative Implications

Preoperative Preparation

- Avoid multiple sticks for IM sedation and IV cannulation.
- Concomitant steroid therapy and the necessity of stress dose should be considered.
- Prophylactic antibiotic therapy because the pt may be on immunosuppressant therapy.
- Prophylaxis against thromboembolism.

Monitoring

- Consider brain function monitoring.
- Arterial catheter as indicated.

Airway

- Oral ulcers, erythema, edema around the lesion, and previous scarring may make airway difficult.
- Use maneuver to reduce pressor response to intubation.
- Avoid airway blocks.

Preinduction/Induction

- Premedication through IV routes.
- Avoid depolarizing muscle relaxant to prevent rise in IOP.

Maintenance

- Depth of anesthesia between 40 and 60 BIS value
- Fluid as per 4:2:1 rule
- Adequate padding of pressure points

Extubation

- Check for any airway trauma or bleeding oral surface.

Postoperative Period

- Pain management by infusion of opioids/suppository

Anticipated Problems/Concerns

- Vasculitis and thrombus formation in larger vessels increases the risk of pulmonary embolism; therefore, initiation of thromboembolic prophylaxis is helpful in such cases.