

Usual Treatment

- Empirical antibiotics (often combination therapy with focus on Gram-positive bacteria) started

after blood cultures obtained when suspicion for IE exists

- Antibiotic treatment for at minimum 4 wk

- Surgery indicated in severe valvular dysfunction, with uncontrolled infection, and to prevent stroke/other embolic process

Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Roth spots (retinal hemorrhage)	Visual disturbance	Retinal exam	Dilated fundoscopic exam
RESP	Pulmonary edema	Dyspnea	Crackles Wheezing	CXR
CV	New or changed murmur Left ventricular or right ventricular failure Conduction abnormality	Dyspnea Orthopnea Light headedness/ syncope	Exam for signs of CHF (left or right sided, depending on lesion)	ECHO (TEE more sensitive than TTE) ECG
GI	Mesenteric ischemia Splenomegaly	N/V Pain	Acute abdomen Hypoactive bowel sounds	Contrasted CT Lactate
CNS	Stroke	Focal neurologic deficit(s)	Detailed neurologic exam	Head CT, MRI
RENAL	Hematuria Glomerulonephritis Pyuria	Urgency Pain Urine discoloration	CVA tenderness	UA
MS	Osler nodes Janeway lesions Splinter hemorrhages	Fever Night sweats Malaise	Skin and nailbed exam	ESR/CRP Rheumatoid factor

Key References: Cahill TJ, Prendergast BD: Infective endocarditis. *Lancet* 387(10021):882–893, 2016; Methangkool E, Howard-Quijano K, Ho JK, et al.: Infective endocarditis: the importance of intraoperative transesophageal echocardiography. *Anesth Analg* 119(1):35–40, 2014.

Perioperative Implications**Preoperative Preparation**

- Assess cardiac status.
- Optimize volume status.
- CT head for baseline study and to rule out embolic stroke.

Monitoring

- Large bore central venous access
- Arterial pressure monitoring
- Noninvasive and invasive cardiac monitoring as indicated

Airway

- Potential for laryngeal involvement (i.e., edema, ulceration, VC paralysis) in pts with SLE.

Induction

- Consider etomidate, ketamine, and/or opioids to avoid SVR reduction.
- Awake arterial line may be beneficial.

Extubation

- Assess and manage postop cardiac dysfunction.
- Consider extubation to NIPPV in pts with continued signs of CHF.

Adjuvants

- Prophylaxis for dental procedures and respiratory tract procedures with biopsy recommended for pts with:
 - Prosthetic heart valves.
 - Unrepaired CHD.
 - Heart transplant with valvular disease.

- Prophylaxis not recommended for bronchoscopy (without biopsy) and GI or GU procedures.

Postoperative Period

- Early postop neurologic exam to assess for CNS embolization

Anticipated Problem/Concerns

- Embolization (intracranial, coronary, mesenteric) can lead to secondary infection.
- Ischemic stroke not a contraindication to surgery, but hemorrhagic stroke requires surgical delay of at least 1 mo.

Epidermolysis Bullosa

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Risk

- 1:17,000, 50% dystrophic form
- Racial distribution equal

Perioperative Risks

- Difficult IV access, airway, intraop positioning, reflux, steroid dependence, intraop hemorrhage, sepsis, iatrogenic corneal abrasion, blister formation, and airway obstruction

Worry About

- Problems similar to those found in pts with severe skin burns; severely compromised pts
- Difficult intubation (23%) secondary to microstomia
- Establishing monitoring and IV access
- Dehydration and malnutrition
- Anemia, hypoalbuminemia, electrolyte imbalance, and thrombocytosis
- Septicemia
- Renal and adrenal dysfunction

Overview

- Characterized by epithelial blistering resulting from minor trauma by lateral shearing forces, not pressure, because of absence of normal intracellular bridges caused by collagen abnormality
- Four types: SEB, JEB, DEB, and Kindler syndrome

- Associated conditions: Growth retardation, pyloric stenosis, esophageal stricture, pseudosyndactyly, enamel hypoplasia, muscular dystrophy, squamous cell carcinoma, and malignant melanoma
- SEB: Most common form; intraepidermal blisters on the soles and palms only in Weber-Cockayne form, generalized in Kobner form, generalized herpetiform in Dowling-Meara form, and generalized in association with muscular dystrophy in the MD form
- JEB: Blisters formed in the intralamina lucida and in intertriginous areas in the inversa form, which are generalized with growth retardation in the Herlitz form, generalized without growth retardation in the non-Herlitz form, and generalized with pyloric atresia
- DEB: Blisters formed in the sublamina densa and in intertriginous areas in the inversa form, on ankles in the pretibial form, on arms and legs in the pruriginous form; generalized blisters in the non-Hallopeau-Siemens form and with growth retardation and severe extracutaneous involvement in the Hallopeau-Siemens form, and aggressive squamous cell carcinomas (very commonly)
- Kindler syndrome: Blisters formed at multiple levels, intralamina lucida and sublamina densa; Kindler syndrome (previously considered as poikilodermatous photosensitivity disease); skin findings including atrophic scarring and nail dystrophy; possibly associated with severe colitis, esophagitis, urethral strictures, and ectropions; squamous cell carcinoma (can develop)

Etiology

- SEB: Inherited autosomal, usually dominant, mutation producing abnormal keratin intermediate filament proteins 5 or 14, which weaken the epidermal architecture; in the MD form abnormality, plectin (cytolinker protein) is the cause.
- JEB: An inherited autosomal recessive mutation produces abnormal laminin 5, abnormal type XVII collagen, and abnormal $\alpha_6\beta_4$ integrin.
- DEB: An inherited autosomal dominant or recessive mutation produces abnormal type VII collagen.

Usual Treatment

- Treatment is supportive, similar to initial burn treatment, with silver impregnated creams and collagen allografts.
- Retinoids and growth-stimulator factors are used to induce wound-repair keratin 6, 16, and 17, which form a more normal epidermis.
- An emerging treatment uses isothiocyanate sulforaphane which induces keratin 16 and 17 and occurs naturally in broccoli sprouts.
- Pts receive steroids and supportive treatment, such as nutritional support, wound care, contracture release, esophageal dilation, oral surgery, and treatment of skin cancers.
- Future treatment is expected to involve gene therapy.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Enamel hypoplasia; Blisters, microstomia, ankyloglossia, supraglottic ulceration or narrowing, corneal erosion, ectropion	Delayed eruption and caries of teeth; painful perioral and intraoral lesions; hoarseness and respiratory obstruction; painful swallowing, spasm, food impaction	Poor oral hygiene and malocclusion; tongue atrophy; obliteration of vestibular sulci, stricture, webs, and vocal cord lesions	Airway assessment, endoscopy
GI	Bullae Perianal blisters, poor absorption, diarrhea	Esophageal stricture Anal pain, tenesmus, constipation	Reflux, regurgitation Anal fissure or stricture	Endoscopy
GU	Blisters	Urinary diversion	Obstruction, sepsis	Renal function
MS	Contractures, growth retardation	Movement limitations, stature	Flexion contracture, pseudosyndactyly	
DERM	Blisters	Age at onset, Hx of remissions and infections	Scars, milia, nail dystrophy, cancer	Skin biopsy

Key References: Lin YC, Goliianu B: Anesthesia and pain management for pediatric patients with dystrophic epidermolysis bullosa, *J Clin Anesth* 18(4):268–271, 2006; Saraf SV, Mandawade NJ, Gore SK, et al.: Epidermolysis bullosa: careful monitoring and no touch principle for anesthesia management, *J Anaesthesiol Clin Pharmacol* 29(3):390–393, 2013.

Perioperative Implications

Preoperative Preparation

- Careful planning of monitoring, IV placement, positioning in the OR, prevention of reflux, and airway management

Monitoring

- No contraindication to pulse oximeter use.
- Protect blisters on the face with foam adhesive inverted to pad mask.
- Pad automated BP cuff heavily and limit intervals.
- Cut off adhesive from ECG leads and hold in place with defibrillator jelly pads.
- Suture invasive monitoring and IVs or wrap in place with petrolatum gauze.
- Esophageal stethoscope may damage mucosa.
- Avoid excessive heat or sweating, which increases the risk of blisters.

Induction

- Regional anesthesia encouraged; use spray antiseptics or pour prep solutions; no intradermal local anesthetics.
- No GA or muscle relaxant specifically contraindicated.

Airway

- All airway management techniques are reported successful.
- The mask (or nasal mask) should be lubricated and padded with petrolatum gauze; pad the chin under fingers; bullae occurred in 1:50.
- LMA one size too small, heavily lubricated, cuff soft with audible leak, extubated deep to prevent trauma; lingual bulla occurred in 1:57.
- Intubation is less frequent; use blind nasal, fiberoptic, and oral techniques; a heavily lubricated small tube and laryngoscope; cricoid pressure without lateral movement is permissible; 66% are class I or II view of larynx and 7–23% have difficult airway incidence; use soft lubricated gauze to prevent tube movement in the mouth; do not allow lateral forces on the corners of the mouth by the tube and do not use tape; the trachea is lined with columnar epithelium and, therefore, is less likely to blister.

Emergence

- Aim for a quiet emergence.
- No suction on intraoral mucosa.

Anticipated Problems/Concerns

- Positioning is performed by the pt if possible; lateral shear forces from lifting cause blisters.
- Corneal abrasion can occur because of poor eyelid retraction; use ointment generously and protect the eyes in while pt is in prone position.
- Treat hemorrhage with epinephrine or a thrombin-soaked sponge.
- Avoid sweating and warming devices, but if unavoidable, the device should be no warmer than skin temperature.
- Extremity tourniquets, IM or rectal medications and EMLA can be used.
- Common procedures include release of syndactyly, dressing change, squamous cell carcinoma, esophageal dilatation, and dental surgery.

Epiglottitis

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Risk

- Prevalent in children 1–7 y; sometimes called supraglottitis, it does occur in adults (decreasing incidence in children >3 y related to vaccines against *Haemophilus influenzae* type B, but still found, particularly if pt is not immunized).
- Adult incidence remains constant with organisms group A *Streptococcus pneumoniae*, *Staphylococcus aureus*, and *Klebsiella pneumoniae*.

Perioperative Risks

- Acute deterioration of airway patency resulting in complete obstruction worse in children
- Difficulty in tracheal intubation due to severe edema of epiglottitis and arytenoids

Worry About

- Airway compromise in children who appear toxic, with increasing distress, drooling, and hypoxemia. The acute risks of airway compromise (of concern

in small children) appear to be less critical in adults, most likely because of larger airways.

- Loss of airway control and aspiration.

Overview

- An acute, potentially life-threatening cause of upper airway obstruction (etiologic agents may include bacteria other than *H. influenzae* type B).
- Produces inflammatory edema of epiglottitis and other supraglottic structures.
- Onset is usually rapid; progression to severe obstruction can occur in several hours.
- High fever, sore throat, and dysphagia are frequently so severe that swallowing is inhibited and drooling results.
- Differential diagnosis also include retropharyngeal abscess (a bacterial infection), which can have the same presentation. It can be differentiated from epiglottitis by the presence of torticollis and trismus and with radiographic studies (contrast CT). Treatment is with antibiotics and surgical drainage.

Etiology

- *H. influenzae* type B is most often traditional associated pathogen, although this can be caused by β -hemolytic streptococci, group A *Streptococcus pneumoniae*, *Staphylococcus aureus*, and *Klebsiella pneumoniae*.

Usual Treatment

- Antibiotic therapy against bacterium (usually *H. influenzae*) and airway support, which generally requires tracheal intubation.
- Because of high incidence of ampicillin-resistant strains, administer ampicillin plus a β -lactamase inhibitor (such as sulbactam) and/or chloramphenicol, cefuroxime, ceftazidime, or another penicillinase-resistant antibiotic as indicated by blood and epiglottitis culture results.
- Tracheal intubation is classically performed in OR in a controlled fashion with surgical support for possible tracheotomy or cricothyrotomy present and gowned.