

- Nearly all affected infants are extubated in OR upon completion of the procedure.
- Transfusion of blood products may be indicated postop.
- Postop hyponatremia is common (31% in one study), although direct complications from hyponatremia seem uncommon.
- Other postop concerns include CSF leak and infections.

Surgical Stages

- A bicoronal incision is made and the scalp and face are dissected to expose the calvarium.
- Calvarial removal may be partial or complete (neurosurgery).
- Supraorbital osteotomies are performed to mobilize the superior orbital rim, nasion, and lateral temporal bones.

- Calvarial vault remodeling and reconstruction with craniofacial plating system is performed.

Anticipated Problems/Concerns

- Difficult airway
- Massive blood loss and transfusion
- Hypothermia
- Possible postop intubation

CREST Syndrome

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Risk

- Pts with Hx of exposure to silica dust or PVC.
- Usual age group is 30–50 y.
- Fourfold to ninefold higher incidence in women than men; seen in all races.
- In USA systemic sclerosis has an estimated incidence of 19 cases per million and prevalence of 240 cases per million (range: 138–286).

Perioperative Risk

- Pts more likely to have compromised renal function at baseline
- Hypoxia from pulm Htn and/or restrictive lung disease
- Difficult intubation from narrow mouth opening

Worry About

- Reflux and thereby aspiration, renal crises, restrictive lung disease, CHF, pulm Htn, difficult intubation due to small mouth opening, and keeping patient warm to avoid Raynaud phenomenon

Overview

- Calcinosis, Raynaud phenomenon, Esophageal dysmotility, Sclerodactyly, and Telangiectasia (CREST).
- Symptoms involved in CREST or limited cutaneous systemic sclerosis are associated with the generalized form of systemic sclerosis.

Etiology

- Exact etiology of systemic sclerosis is unknown; the following pathogenic factors are always present: Endothelial cell injury, fibroblast activation, and cellular and humoral immunologic derangement.
- Environmental factors, such as silica, industrial solvents, and radiation exposure, are all triggers or accelerators.
- CMV, HHV5, and parvovirus are possible viral accelerators.

Usual Treatment

- Glucocorticosteroid, immunosuppressive, chelating agents, endothelin receptor antagonist, PDE5 inhibitor, and peripheral vasodilators.

- Skin thickening with D-penicillamine and γ -interferon (not FDA approved).
- Pruritus with moisturizers, camphor and menthol, H₁ and H₂ blockers, tricyclic antidepressants, PUVA, UVA-1 phototherapy, and trazodone.
- Raynaud phenomenon with CCB, prazosin, PGE1, dipyridamole, aspirin, smoking cessation, and topical nitrates.
- GI symptoms with antacids, H₂ blockers, PPIs, prokinetic agents, and octreotide.
- Lung symptoms with CCB, PGs, cyclophosphamide, and high-dose corticosteroids.
- Renal involvement with ACE or angiotensin II inhibitors.
- Cutaneous telangiectasias can be treated by PDL and IPL; PDL has better cosmesis but more side effects.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
DERM/HEENT	Sclerodactyly, few wrinkles or joint creases, decreased range of motion, hair loss, pruritus telangiectasies	Observation	Tightness, indurations, hyperpigmentation or hypopigmentation	Airway examination
CV	Pericardial effusion, CHF, myocardial fibrosis, misconduction Cor pulmonale	Dyspnea, palpitation, irregular heart rate, chest pain from vasospasm	Rales and murmurs on auscultation	ECG, Holter monitoring, ECHO
RESP	Pulm Htn, aspiration pneumonia, dyspnea	SOB, cough, tachypnea, dec exercise tolerance	Dry rales	PFT, ABG, CXR, DLCO, HRCT
CNS	Carpal tunnel syndrome, trigeminal neuralgia (rare), entrapment neuropathies	Pain over wrist, other typical signs depending on nerve involved	Limited ROM	Conduction studies, CT
RENAL	Htn, oliguria	Headache, SOB, edema	Swelling of hands and feet	Check BP and UO and monitor serum Cr
MS	Raynaud phenomenon, arthralgias, myalgias, morning stiffness	Acroosteolysis, muscle weakness	Palpable tendon friction rubs, muscle wasting, flexion contractures	Increased serum CK and aldolase
GI	GE reflux, esophagitis, esophageal strictures, watermelon stomach, primary biliary cirrhosis, colonic diverticula, anal sphincter incompetence	Bitter taste, dysphagia, retrosternal and abdominal pain, diarrhea, self-soiling	Abdominal tenderness, decreased rectal sphincter tone	Barium swallow CT or MRI, endoscopy Abdominal ultrasound, antimitochondrial antibodies for PBC

Key References: Gabrielli A, Awedimento EV, Krieg T. Scleroderma. *N Engl J Med* 360(19):1989–2003, 2009; Pritts CD, Pearl RG: Anesthesia for patients with pulmonary hypertension, *Curr Opin Anaesthesiol* 23(3):411–416, 2010.

Perioperative Implications

Preoperative Preparation

- Continue PPI, consider FOI, and evaluate for regional anesthetic techniques for pulm issues.

Monitoring

- If comorbidities dictate, arterial line (try to avoid due to Raynaud phenomenon, but difficult to get cuff pressure due to reduced flow; may need ABG)
- CVP \pm PA cath if pulm Htn, along with standard monitoring

Airway

- Airway may be a challenge due to small oral opening,

Preinduction and Induction

- Worry about hypotension and hypoxemia at induction,

Maintenance

- Choose drugs based on hemodynamic status,
- Keep warm,

Extubation

- May have to be delayed if significant pulm compromise

Adjuvants

- In the presence of compromised renal, cardiac, or pulm function, modify anesthetic drugs accordingly

Anticipated Problems/Concerns

- Challenging airway, hypoxemia, CHF, renal function, and positioning challenges with contractures