Usual Treatment

- Parity of RCTs; combination of therapies
  - Plasmapheresis (exchange): Daily, more NB than plasma infusion (platelet-poor FFP) usually with rapid clinical response in days (failed Rx within 4–7 d)
  - Steroids: Adjuvant (methylprednisolone 10 mg/kg/d better than 1 mg/kg/d). Potentially suppress production of anti-ADAMTS13 autoantibodies.
  - Rituximab: Monoclonal Ab against CD20 Ag on B lymphocytes; used to treat refractory or relapsing TTP. Optimal dosing, timing, and side effects based on case series.
  - Splenectomy: For failed medical response, prevention of relapse (small series).
  - Avoid platelet transfusion.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Airway manipulation, potential hemorrhage			
CV	Rare conduction pathway involvement		Baseline MAP for perfusion CNS/kidney Vascular access	ECG
RESP	Rare infiltrates causing hypoxemia			CXR
RENAL	Proteinuria, hematuria, ARF less common than HUS			BUN, serum Cr, urine sediment
HEME	Thrombocytopenia	Hemorrhage	Petechiae Jaundice	PT, PTT, fibrinogen usually normal (differentiate from DIC). Fragmented RBCs (anemia); increased LDH, bilirubin, decreased haptoglobin
CNS	Fluctuating course	Spectrum—headache, seizures, coma		Lumbar puncture, EEG, neuroradiology studies
OB	May precipitate episode or relapse	Differentiate from HELLP/PIH (more CNS, less hepatic involvement, not improved postpartum)		

Key References: Sayani FA, Abrams CS: How I treat refractory TTP, *Blood* 125(25):3860–3867, 2015; Weinberg L, Chang J, Hayward P, et al.: Post-cardiac surgery TTP with digital ischemia, *Anaesth Intensive Care* 41(3):386–389, 2013; Pourrat O, Coudroy R, Pierre F: Differentiation between severe HELLP syndrome and thrombotic microangiopathy, TTP and other imitators, *Eur J Obstet Gybecol Reprod Biol* 189:68–72, 2015.

Perioperative Implications

Preoperative Preparation

- Steroid supplement if receiving.
- Premedication: Not IM; caution with CNS involvement.
- Pneumococcal, meningococcal (*Haemophilus influenzae* for children) if splenectomy is anticipated.

Monitoring

- Protect skin, mucous membranes (NIBP cuff, esophageal probe, pressure points)
- Central access is usually provided for plasma exchange. If this is required, avoid subclavian vein if possible (difficulty compressing hematoma).
- Theoretical risk of radial arterial line with thrombotic process.

Airway

- Avoid nasal ETT. Be careful with instrumentation, especially if platelet count $<50{,}000 \times 10^3/mm^3$.

Induction

- Avoid sympathetic intubation response (CNS disease spectrum), maintain MAP > CNS, renal autoregulatory thresholds (>50–60 mm Hg).

Maintenance

- Theoretical advantage of inhibitory effect volatile anesthetics on platelet aggregation

Fluids

- Do not transfuse platelet unless there is life-threatening thrombocytopenia: There are reports of deterioration due to further microthrombi.
- Bleeding managed with RBCs (>48 h old to avoid active platelets) and FFP (platelet-poor).

Extubation

- As above: Care of mucous membranes and hemodynamic response

Adjuvants

- Individual analysis of risk/benefit for neuraxial technique in thrombocytopenic pt (if in remission)

Postoperative Period

- Mobilize early: Precipitous increase in platelet count and viscosity with risk of thrombotic events.

Anticipated Problems/Concerns

- Hemorrhage if there is life-threatening thrombocytopenia (no platelet transfusion until then)
- Microthrombi with CNS dysfunction

Pyloric Stenosis

Inna Maranets

Risk

- Incidence: 1:300-1000 live births.
- Incidence is 3–5% higher among children of affected parents.
- More common in males.

Perioperative Risks

- Similar to other abd procedures in pts of same age.
- Some association with GU anomalies.
- Some pts have elevated unconjugated bilirubin related to decreased glucuronyl transferase activity; this returns to normal after correction of stenosis.

Worry About

- Full stomach. Recurrent emesis leads to dehydration, electrolyte imbalance, and alkalosis.
- Typically pts have hypochloremic, hyperkalemic metabolic alkalosis.
- Metabolic acidosis found in the most severe cases.

Overview

- Reduced size of gastric outlet impedes emptying of contents, which can cause abnormal nutrition, gastric distention, repeated vomiting, and dehydration.
- Onset of symptoms occurs at 3–6 wk of age.
- Usually surgically cured.

Etiology

- Almost exclusively genetic in infants
- Can be acquired in adults

Usual Treatment

- Normalize fluid/lyte status: This is not a surgical emergency.
- Surgical: Pyloromyotomy can usually be undertaken within 2–24 h of admission (unless fluid derangements are severe).
- Short procedure (<1 h): Open or laparoscopic.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
GI	Gastric outlet obstruction	Nonbilious projectile emesis	Pyloric "olive" palpable in upper abdomen	Contrast study Abdominal US

Key References: Schapiro F, Litman RS: Pyloromyotomy. In Litman RS, editor: *Pediatric anesthesia practice*, New York, NY, 2007, Cambridge University Press, pp 173–175; Considine AA, Maranets I, Snegovskikh D, et al.: Induction and airway management for pyloromyotomy, *J Anesth Clinic Res* S3:003, 2011.

Perioperative Implications

Preoperative Preparation
- Correct fluid and acid-base deficits.
- Place orogastric tube to aspirate contents.
- Pyloric stenosis is not a surgical emergency.

Monitoring
- Routine

Airway
- Full stomach

Preinduction/Induction
- IV atropine 0.02 mg/kg; minimal dose 0.1 mg.
- Empty stomach with orogastric tube or suction cath.
- Consider awake intubation or IV RSI, especially if pt received barium contrast.
- Modified RSI with either propofol alone or propofol and nondepolarizing muscle relaxant; cricoid pressure and mask ventilation have been used successfully without increased incidence of complications.
- Hypoxemia is common during rapid-sequence induction; ventilate with cricoid pressure.
- In a properly resuscitated pt with recent loss of IV access, inhalational induction has been used successfully and can be considered a safe alternative.

Maintenance
- No technique is absolutely contraindicated by pyloric stenosis alone.
- Inhalational agent in O_2 and air or N_2O, short or intermediate-acting muscle relaxant.
- Avoid opioids.
- Local infiltration with bupivacaine or ropivacaine by surgeon.
- IV fluids should be warmed.
- Replacement fluids: LR 1–2 mL/kg per h.
- May consider using D_5 if the procedure lasts more than 1 h.

Extubation
- Potential of full stomach; suction stomach prior to extubation.
- Reverse NMB agent.
- Awake extubation
- Delayed awakening is common

Adjuvants
- Consider potential of associated liver and GU abnormality.

Postoperative Period
- Potential for central apnea and reactive hypoglycemia
- Pulse oximetry/apnea monitoring for the first 12–24 h
- Continue IV glucose until there is adequate PO intake.
- Pain score: 2–5, acetaminophen is usually sufficient; avoid opioids.

Anticipated Problems/Concerns
- Potential for full stomach.
- Need to correct fluid and/or electrolyte imbalances preop.
- Delayed awakening is common.

Q Fever

Thomas A. Russo | Paul R. Knight III

Risk
- Greatest after direct or indirect exposure to infected cattle, sheep, or goats; particularly at parturition
- Less from a variety of other animals, rarely from blood products
- Abattoir workers, veterinarians, and other animal workers at greatest risk
- Pts with immune impairment are at a higher risk (e.g., HIV, steroids)
- Mortality 2.4% overall; chronic infection ~16%.

Perioperative Risks
- Decreased respiratory reserve secondary to pneumonia
- Decreased myocardial reserve secondary to endocarditis
- Further increase in hepatocellular injury if there is liver involvement

Worry About
- Secondary respiratory complications
- Decreased myocardial performance and emboli with endocarditis
- Hepatic or neurologic involvement

Overview
- Acute infection: Asymptomatic (~50%) to moderate severity (2% hospitalized).
- Acute symptomatic disease presents as nonspecific febrile syndrome ± pneumonitis (~50%), hepatitis (80% or more), pericarditis and/or myocarditis (<5%), neurologic disease (<5%).
- Chronic disease occurs in <1% of infections, usually without fever.
- Chronic disease, primarily endocarditis (particularly abn or prosthetic valves) and occasionally bone.

Etiology
- *Coxiella burnetii*, the causative organism, is a fastidious obligate intracellular bacterium.
- The spore stage can withstand harsh environmental conditions for prolonged periods, facilitating indirect transmission.
- Highly infectious; transmitted (1–10 organisms) primarily by inhalation, from unpasteurized milk, or by a tick bite.
- Incubation period ~20 d (range, 3–40 d).
- Bacterium targets reticuloendothelial cells and develops into granuloma.

Usual Treatment
- Dx: Epidemiologic circumstance and serology (positive in 2–4 wk).
- Acute disease: Doxycycline or quinolones for 2–3 wk hastens resolution.
- Chronic disease: Doxycycline and rifampin for 1–3 y; with endocarditis, possible valve replacement.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
CV	Endocarditis Immune complex vasculitis Microthromboembolism	Rash, reduced exercise tolerance	Clubbing, rash, murmurs, petechiae	ECHO, ECG, culture negative with standard techniques serology, PCR
RESP	Atypical pneumonia, asymptomatic pneumonia, rapidly progressive pneumonia, interstitial pulm fibrosis	Pleuritic chest pain, cough, dyspnea	Consolidation, rales, pleural effusions	CXR, serology
GI	Acute hepatitis	N/V, fatigue, diarrhea, sweats and chills	Hepatomegaly or hepatosplenomegaly	SGOT, SGPT, bilirubin, granulomas on liver biopsy
HEME	Hyperglobulinemia, anemia, thrombocytosis/thrombocytopenia	Easy fatigue, bleeding tendency	Pallor; purpuric eruptions	Sedimentation rate, Hct/Hgb, plt count
OB	Immune complex vasculitis Q fever complications secondary to reactivation of infection during pregnancy	Spontaneous abortion more likely		Microscopic hematuria Isolation of *C. burnetii* from placenta
CNS	Meningoencephalitis Optic neuritis	Weakness, seizures, meningismus, blurred vision, headache	Focal deficits, sensory loss	Increased monocytes and protein in CSF; normal glucose
MS	Immune complex vasculitis, vertebral osteomyelitis	Myalgia	Point tenderness	X-ray

Key References: Marrie TJ: *Coxiella burnetii* (Q fever). In Mandell GL, Bennett JE, Dolin R, editors: *Mandell, Douglas, and Bennett's principles and practice of infectious diseases*, ed 5, New York, NY, 2000, Churchill Livingstone, pp 2043–2050; Schaik EJ, Chen C, Mertens K, et al.: Molecular pathogenesis of the obligate intracellular bacterium *Coxiella burnetii*, *Nat Rev Microbiol* 11(8):561–573, 2013; Eldin C, Melenotte C, Mediannikov O, et al.: From Q fever to *Coxiella burnetii* infection: a paradigm change, *Clin Microbiol Rev* 30:115–190, 2016.