with the population that does receive autologous blood transfusion.

Worry About

- Understanding the rights and desires of pt versus duty of physician in regard to blood or blood product administration.
- Trauma and emergency situations in which little time is available to discuss blood product transfusion.
- Competent adults are those who know the nature and consequences of their actions and such adults have the right to refuse specific therapies.
- *Parens patriae* ("parent of the nation") refers to the public policy power of the state and represents the duty and interest of the state to preserve the health of minors. Medicolegally, when a child's right to live and parental religious beliefs collide, the courts have consistently ruled that the child's welfare is paramount.

Overview

- Began as Bible study group in 1869 and adopted the name Jehovah's Witnesses (based on Isaiah 43:10–12) in 1931.
- Strict interpretation and adherence to Biblical passages, which forbid eating of blood. This is interpreted as prohibition of acceptance of blood products to sustain life because this may compromise their soul.
- Other medical restrictions were established over time, such as prohibition of organ transplants in 1967. However, vaccinations are deemed acceptable.
- In 1942 the Watchtower Society, the governing body of Jehovah's Witnesses, introduced the blood ban, which forbids members from accepting allogeneic blood products, including whole blood, RBCs, WBCs, platelets, and plasma.
- There is variability among members to the interpretation of the prohibition regarding blood. Jehovah's Witnesses may consider the use of one's own blood in the course of a medical procedure or therapy provided there is no advanced storage. They may accept fractions of plasma, such as albumin, rHuEpo, immunoglobulin, or factor concentrates.

Usual Treatment

- Discuss and document preoperatively the potential for life-threatening hemorrhage. Discuss and document therapies and interventions that would be acceptable to the pt.
- Seek evidence of an advance directive, an affidavit that confirms the pt's refusal to accept a transfusion (which promotes discussion and releases physicians/hospitals of responsibility for outcome of the pt's decision).
- Consider contacting a Jehovah's Witness Hospital Liaison Committee, which consists of a group of individuals trained to work as intermediaries in avoiding conflict between pts and physicians.
- Contact legal counsel if pt is a minor, unconscious, or an incompetent adult.
- Be aware that administration of blood products against a competent pt's wishes can be a prosecutable offense.

Assessment Points

System	Assessment by Hx	Test
HEME	Evaluate for treatable forms of anemia	Hg/Hct, folate, B_{12} levels, Fe, ferritin, transferrin saturation

Key References: Bodnaruk ZM, Wong CJ, Thomas MJ: Meeting the clinical challenge of care for Jehovah's witnesses, *Transfus Med Rev* 18(2):105–116, 2004; Lawson T, Ralph C: Perioperative Jehovah's witnesses: a review, *Br J Anaesth* 115(5):676–687, 2015.

Perioperative Implications

Preoperative Preparation

- Iron therapy, especially if evidence of decreased iron stores: Ferrous sulfate 325 mg PO daily or iron dextran 100–200 mg IV daily.
- Vitamin B_{12} 1 mg IV once daily.
- Folate 1-5 mg IV daily.
- Consider rHuEpo: 600 U/kg SQ for 21 d prior to surgery.
- Delay elective surgery until red cell mass is optimal.
- Consider anesthetic alternatives such as regional or neuraxial anesthesia.

Monitoring

- Minimize phlebotomies. Consider pediatric sampling tubes.
- Consider central venous line, pulm artery cath, and arterial line if high possibility of hemorrhage.

Intraoperative Considerations

Maintain Blood Volume

- Nonblood volume expanders (i.e., normal saline, lactated Ringer, PlasmaLyte A, hydroxyethyl starches, dextrans).
- Synthetic oxygen therapeutics (recombinant human hemoglobin).
- Hypervolemic or normovolemic hemodilution (maintain continuous circuit with pt) in the absence of CAD or Hg <7 g/dL.
- Blood salvage techniques (maintain continuous series with pt's circulation).
- Red cell substitutes include crystalloids, colloids, recombinant erythropoietin, and recombinant factor VIIa; in some cases human, animal, or synthetic hemoglobin may be acceptable.
- White cell substitutes include interferons and interleukins and should be considered on a case by case basis.
- Plasma may be substituted with albumin, immunoglobulin, cryoprecipitate, and/or clotting factors. Determine if acceptable with the pt before administering.

Maximize Oxygen Delivery

- Increase FIO_2.
- Hyperbaric O_2.
- Inotropic agents to augment cardiac index once volume resuscitated.

Prevention of Intraoperative Blood Loss

- Meticulous surgical technique and use of hemostatic surgical instruments.
- Avoiding blood loss is most effective in preventing mortality.
- Consider use of tourniquet if feasible for particular surgery.
- Consider use of antishock garments, such as pneumatic dressing.
- Laparoscopic, endovascular, or minimally invasive surgical techniques.
- Hypotensive anesthetic techniques.
- Preoperative angiographic embolization (i.e., uterine arteries for hysterectomy).
- Correct coagulopathies with pharmacologic agents (tranexamic acid, aminocaproic acid, desmopressin, recombinant factor VIIa).
- Hemostatic products containing blood fractions (fibrin glue/sealant, thrombin sealants)

Minimize O_2 Consumption and Demand

- Hypothermia 30–32° C (reduces O_2 consumption 50%); however, also a concern for hypothermia induced coagulopathy. Consider risks and benefits.
- Sedation, analgesia, paralysis.
- Acute hypervolemic hemodilution, CPB or ECMO, renal dialysis, and plasmapheresis.

Postoperative Considerations

- Consider postop ventilation with paralysis, sedation, and hypothermia for severe anemia.
- Consider PA catheter to measure and follow CO and SvO_2 to assess O_2 delivery and consumption without resorting to phlebotomy.
- Supplement with IV hyperalimentation, rHuEpo, and iron dextran.
- Avoid gastric ulceration with proton pump inhibitors.
- Consider progesterone for control in menstrual bleeding.

Jeune Syndrome (Asphyxiating Thoracic Dystrophy)

Anne M. Lynn | K. Karisa Walker

Risk

- Incidence in USA: 1:100,000-130,000 live births and prevalence of 2.6:100,000.
- No race or sex predilection.
- Skeletal survey by US after 14 wk gestation can detect defining deformities.
- Heterogeneous presentation, from mild to fatal.

Perioperative Risks

- 70–80% mortality in infancy for severe cases
- Respiratory failure from small thoracic cage and hypoplastic lungs; frequent infection in those with ciliary dysmotility
- Progressive renal disease with cystic lesions and periglomerular fibrosis
- Liver and pancreatic involvement with fibrosis and cysts

Worry About

- Hypoxic and/or hypercapnic respiratory failure.
- Barotrauma with positive pressure ventilation.
- Renal failure requiring careful fluid and electrolyte management and selection of nonrenally cleared muscle relaxants and opiates.
- Liver involvement, and rarely cirrhosis, may affect drug metabolism.

Overview

- Rare autosomal recessive disease with skeletal dysplasia and variable renal, hepatic, and eye abnormalities; variable involvement of CNS and GI systems.
- Overlap of findings with Ellis-van Creveld syndrome, short rib polydactyly syndrome, and oral-facial-digital syndromes.
- Poor survival beyond early infancy.
- Narrow, rigid thoracic cage due to short horizontal ribs, short limbs, underdeveloped iliac wings and acetabula and occasional polydactyly.
- Respiratory failure from restrictive thorax and hypoplastic lungs.
- Possible renal, hepatic, and pancreatic dysfunction if pt survives infancy.
- Chronic renal failure can lead to transplantation.
- Hepatic dysfunction can be controlled with ursodeoxycholic acid but those with severe portal Htn require liver transplantation.
- Occasional pulm Htn and cor pulmonale.
- Surgical enlargement of the thorax has been undertaken to increase chest wall compliance.

Etiology

- Autosomal recessive inheritance, variable phenotype
- Postulated involvement of chromosome 15q13 or *IFT80* gene on chromosome 3

Usual Treatment

- VEPTR thoracoplasty and external distraction thoracoplasty have been successful for Jeune syndrome.
- Older children may require surgery related to renal or hepatic failure or treatment of retinal pathology.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
CV	Pulm Htn	Syncope	Increased second heart sound	ECG (RVH) ECHO
RESP	Stiff, small rib cage Hypoplastic lungs Ciliary dysfunction	Pneumonia/respiratory failure Assisted ventilation Asynchronous ventilation with agitation/crying	Small chest Horizontal ribs Cyanosis with crying	ABG CXR Oximetry
GI	Hepatic fibrosis/cysts Pancreatic fibrosis/cysts Foregut dysmotility/malrotation	Failure to thrive Metabolic anomalies Nausea, vomiting	Hepatomegaly	Abdominal US Bilirubin/ LFTs
RENAL	Cysts Nephritis	Polyuria, polydipsia		BUN, Cr, lytes, Ca^{2+}/PO_4, abdominal US
CNS	Occasional hydrocephalus Dandy-Walker malformation Retinal degeneration		Increased OFC (head circumference)	
MS	Short limbs and stature Polydactyly of hands and feet			X-ray of thorax, pelvis

Key References: Keppler-Noreuil KM, Adam MP, Welch J, et al.: Clinical insights gained from eight new cases and review of reported cases with Jeune syndrome (asphyxiating thoracic dystrophy), *Am J Med Genet A* 155A(5):1021–1032, 2011; Waldhausen JH, Redding GJ, Song KM. Vertical expandable prosthetic titanium rib for thoracic insufficiency syndrome: A new method to treat an old problem, *J Pediatr Surg* 42:76-80, 2007.

Perioperative Implications

Preoperative Preparation

- Assess ventilation.
- Evaluate for possible pulm Htn.
- Evaluate renal function and consider LFT.
- High index of suspicion for other organ system involvement.

Monitoring

- Consider arterial catheter.
- Consider central venous access.

Airway

- Small larynx requires smaller ETT size.

Induction

- Agitation may make respirations asynchronous (chest and/or abdomen), causing hypoxemia.

Maintenance

- Lung hypoplasia makes barotrauma high risk; maintain low peak airway pressures.

Extubation

- Ensure adequate ventilation before extubation; postop ventilation may be needed for a prolonged period, specifically after thoracoplasty.

Adjuvants

- Renal/hepatic function assessment guides selection of muscle relaxant and fluid management.

Anticipated Problems/Concerns

- Asynchronous ventilation during crying with hypoxia
- Barotrauma during assisted mechanical ventilation
- Renal and/or liver disease and drug metabolism
- Postoperative respiratory failure requiring ventilatory support

Juvenile Gaucher Disease (Type III/Subacute Neuronopathic)

Lance C. Atchley | Lee A. Fleisher

Risk

- Less than 1:100,000
- Autosomal recessive with no sex predominance
- Panethnic but more common in Northern Sweden and Palestinian town of Jenin

Perioperative Risks

- Abnormal platelet functioning and increased risk of bleeding
- Respiratory failure
- Seizure

Worry About

- Intraoperative blood loss and need for transfusion of PRBC, FFP, and platelets
- Perioperative continuation of anticonvulsant therapy and possible need to supplement
- GERD and aspiration
- Potential presence of restrictive or obstructive lung pathology
- Potential presence of pathologic fractures, including vertebrae
- Type IIIc disease: Intracardiac calcifications—mitral valve, aortic valve, ascending aorta, aortic arch, and coronary ostia

Overview

- Variable clinical expression and severity; presents along a continuum.
- Systemic involvement often present in all forms of Gaucher disease, including type III:
  - Splenomegaly, which may lead to anemia, thrombocytopenia, and leukopenia.
  - Platelet dysfunction independent of splenic involvement.
  - Decrease in coagulation factors.
  - Hepatomegaly.
  - Skeletal involvement, including bone marrow infiltration, osteonecrosis/osteoporosis, and pathologic fractures.
- Systemic involvement more common in type III disease:
  - Pulm involvement ,including interstitial lung disease, pulm Htn, or hepatopulmonary syndrome.
  - GERD with chronic aspiration.
- Specific to type III disease:
  - Slowly progressive neurologic symptoms.
  - Supranuclear horizontal gaze palsy is pathognomonic sign.
  - Seizures may be present.
  - Oculomotor apraxia.
  - Three subtypes:
    - Type IIIa: Myoclonus; dementia