

- Neonate: Physiologic jaundice of the newborn, breast milk jaundice, hypothyroidism, galactosemia
- Sepsis
- Acquired transferase deficiency: Drug inhibition (e.g., pregnanediol, chloramphenicol), hepatocellular disease (cirrhosis, hepatitis)
- Gilbert disease: Decreased glucuronyl transferase; usually mild but can transiently worsen during periods of stress
- Crigler-Najjar I (absent) and II (partial decrease) in glucuronyl transferase
- Mostly conjugated-decreased hepatic/extrahepatic excretion:
 - Hereditary and/or familial: Dubin-Johnson, Rotor syndromes, recurrent intrahepatic cholestasis

- (benign), gestational cholestatic jaundice (approximately 1:13,000 deliveries; third trimester; pre-eclampsia, nulliparity; twin; decreased plt)
- Acquired: Sepsis; hepatocellular disease (drug- and viral-induced hepatitis), postoperative jaundice (pigment overload [transfusions, resorption of hematomas, hemolysis]), hepatocellular damage [drugs, including halothane; shock], benign postoperative jaundice, drug-induced cholestasis (e.g., oral contraceptives, methyltestosterone)
 - Extrahepatic biliary obstruction (e.g., mechanical, from stones, stricture, tumor, pancreatitis)
- Pseudojaundice:
 - Dietary carotenoids (primarily infants; excessive intake of vegetables, such as

- carrots and tomatoes), TPN-associated liver dysfunction
- Poisoning (picric acid)

Usual Treatment

- No specific treatment outside of newborn period.
- For neonates: fluids, phototherapy, exchange transfusion, albumin, tin mesoporphyrin, and IV immunoglobulin Rx have been shown to decrease the level of unconjugated bilirubin below levels regarded to be toxic to the neonatal brain. The smaller and sicker the premature infant, the more aggressive the therapy needed.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT		Duration	Yellow sclerae	
CV	Hyperdynamic Poss decreased SVR	General symptoms	Increased HR, decreased BP	
RESP	Cirrhotics have 6x increase in pulm Htn	Severe dyspnea, hypoxia, clubbing	Clubbing Cyanosis	ECHO; right-sided heart cath if indicated, usually for PAS ≥50
GI	Severe dysfunction Prolonged effects of most anesthesia drugs	General symptoms, reflux, ascites, varices, edema	Signs of chronic liver disease	LFTs Coagulation time Hgb, plt
ENDO/METAB	Decreased synthetic function, increased enzymes, decreases albumin, decreased hepatic coagulation factors; decreased clearance of toxins	General malaise symptoms Easy bruising and bleeding	Jaundice Ecchymoses Hematoma Ascites	LFTs Coagulation time NH ₃ , lactate
HEME	Decreased plt	Easy bruising and bleeding	Ecchymoses, hematoma	
DERM		Duration, evidence of bleeding	Yellow color	
RENAL	Decreased function Higher risk of postop renal impairment		Edema May be hypovolemic in obstructive jaundice	BUN, Cr; Cr may be spuriously lower with high bilirubin
CNS	Recurrent encephalopathy in cirrhosis Cerebral edema in fulminant hepatic failure Autonomic dysfunction	Mental status Duration of illness Abnormal autonomic function	Normal to encephalopathy/ comatose Orthostatic BP changes	Bilirubin interferes with cerebral near-infrared oximetry

Key References: Yang LQ, Song JC, Irwin MG, et al.: A clinical prospective comparison of anesthetics sensitivity and hemodynamic effect among patients with or without obstructive jaundice, *Acta Anaesthesiol Scand* 54(7):871-877, 2010; Vaja R, Barker RC: Drugs and the liver, *Anaesth Intensive Care Med* 13:71-74, 2011.

Perioperative Implications

- Drug: Decreased protein production leads to decreased albumin binding and more active drug.
 - Cimetidine and/or ranitidine: Clearance reduced, especially in pts with ascites, hypoproteinemia, and encephalopathy.
 - Benzodiazepines: Clearance of oxidative pathway markedly decreased; glucuronidation path (e.g., lorazepam) not greatly altered; excessive sedation in severe liver disease.
 - Narcotics: Meperidine clearance is severely affected; adverse affects of morphine can be increased.
 - Neuromuscular blockade: Succinylcholine activity may be prolonged somewhat because of decreased levels of pseudocholinesterase; decreased metabolism of vecuronium and rocuronium.
 - Miscellaneous: Phenobarbital and lidocaine have reduced clearance; diuretics may have reduced natriuretic efficacy.

- Halogenated agents: Halothane should be avoided; association of enflurane with hepatic toxicity is less clear; isoflurane and sevoflurane are preferred agents in setting of liver disease and best preserves liver hemodynamics; reports of hepatic toxicity for both are rare.
- Pregnancy: Jaundice may signal HELLP syndrome and pregnancy-induced Htn.
- Cardiac surgery: Jaundice occurs in approximately 20% post-CPB pts; risk factor for mortality.

Preoperative Preparation

- Hydration should be adequate; if chronic liver failure, may be total body fluid increased but intravascularly decreased.

Monitoring

- NMB: Dose muscle relaxants to effect and consider path of elimination.
- Invasive CV monitoring: Important for some procedures.

Airway

- May have bleeding disorder.

Induction

- Avoid benzodiazepines.
- Consider cricoid pressure if varices present.

Maintenance

- Be mindful of metabolic clearance paths.
- When practical, use drugs cleared chiefly by nonhepatic paths.

Extubation

- May have delay in awakening

Anticipated Problems/Concerns

- Inability to extubate immediately postoperatively due to prolonged action of NMB and sedative/hypnotic/narcotic medications

Jehovah's Witness Patient

Meg A. Rosenblatt | Aloi Patel

Risk

- More than 8 million members worldwide
- Headquarters in Brooklyn, New York; new world headquarters under construction in Warwick, New York

Perioperative Risks

- Possible morbidity and/or mortality from massive hemorrhage secondary to religious dogma banning members from accepting blood transfusions.

- Appropriate blood conservation measures (i.e., autologous blood salvage, normovolemic hemodilution, reduction of intraoperative and iatrogenic blood loss) in pts who do not accept autologous blood transfusions results in similar or better outcomes compared

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.

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 B₁₂ 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.

- 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.

- 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 FIO₂.
- Hyperbaric O₂.
- 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 O₂ Consumption and Demand

- Hypothermia 30–32° C (reduces O₂ 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 SvO₂ to assess O₂ 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.