Anemia, Hemolytic

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Risk

- Autoimmune disorders (SLE, RA, scleroderma, cold agglutinin disease).
- Lymphoproliferative disorders (CLL, NHL).
- Prosthetic heart valves (ball-and-cage, and bileaflet valves). Usually subclinical, but can be severe in up to 15% of pts.
- Family history of hemoglobinopathies or RBC membrane defects (thalassemia, sickle cell disease, G6PD deficiency, spherocytosis).
- Exposure to drugs (cephalosporins, penicillins, NSAIDs) or other chemicals (naphthalene, fava beans).
- Infection (*Clostridium perfringens*, *Haemophilus influenza* type B, malaria, HIV).
- Wilson disease (due to toxic effect of copper ions in circulation).

Perioperative Risks

- Anemia, hypoxia.
- Underlying CV compromise.
- Splenomegaly in pts with extravascular hemolysis (within the reticuloendothelial system). Splenectomy is a common surgical procedure in pts with sickle cell disease due to hemolysis and sickling.
- Renal failure due to massive hemolysis (e.g., cold agglutinin hemolysis, sickling, drug reaction)
- Varying levels of liver disease depending on type of hemolytic anemia. Synthetic function of liver is usually normal, but in severe cases can be compromised.

Worry About

- Uncompensated anemia in pts with subacute hemolysis
- Periop hemolysis and/or hypoxia
- Need for transfusion and/or fluids

Overview

- Pts with hemolytic anemia may present with any of the following: fatigue, angina, SOB, tachypnea, tachycardia, or jaundice. The hemolysis can lead to changes in blood viscosity, gallstone production, splenomegaly, and renal failure in severe cases. Many pts will be both iron and folate deficient.
- Epidemiology varies by pt population. For example, G6PD is an X-linked condition and its prevalence is near 50% in Kurdish Jews, but around 1:1000 in North American and European populations.
- Other things to consider incl monitoring periodic Hct levels, and administering prophylactic antibiotics/vaccinations to pts who have had a splenectomy.

Etiology

- Multiple causes; see Risk section (e.g., RBC structural abnormalities, autoimmune reaction, enzyme deficiency, hemoglobinopathies, mechanical heart valves, drugs, infection).

Usual Treatment

- Treatment depends on etiology:
  - Autoimmune: Corticosteroids, plasmapheresis, packed RBC transfusion for symptomatic pts, supportive care;
  - Drug induced: Discontinuation of offending medication, corticosteroids, supportive care;
  - Prosthetic valve: Cardiology consult and transfusion if symptoms rapidly worsen;
  - RBC membrane defect: Splenectomy and supportive care;
  - Enzyme deficiency: Avoidance of triggers, splenectomy, supportive care;
  - Infection: Treatment of underlying infection and supportive care;
  - Wilson disease: Rapid removal of copper, early consideration for liver transplant.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
CV	Dehydration	Fatigue, dizziness	Hypotension, weak pulses, increased capillary refill	CBC, BNP
HEME	Anemia	Fatigue, SOB, dizziness	Jaundice, pallor, splenomegaly	Hgb, Hct, reticulocyte count, indirect bilirubin, LDH
RENAL	Hemoglobinuria, acute renal failure	Dark urine (episodic)	Possible Htn, resp rate changes	Urine analysis, BUN, Cr
GI	Liver disease		Hepatosplenomegaly	Liver function tests (LFTs)

Key Reference: Eckman JR: Disorders of red cells. In Lubin MF, Dodson TF, Winawer NH, editors: *Medical management of the surgical patient: a textbook of perioperative medicine,* ed 5, Cambridge, 2013, Cambridge University Press, p 215.

Perioperative Implications

Preinduction/Induction/Maintenance

- Preop management and treatment of underlying cause of hemolytic anemia.
- The test obtained periop depends on the etiology, severity, and chronicity of the hemolytic anemia.
- Avoidance of hypoxia, hypercarbia, acidosis, low-flow conditions, and hypothermia (particularly in cold agglutinin disease).
- Optimize CV status with adequate hydration; consider IV fluid treatment the day before surgery if hypovolemic.
- RBC transfusion may be considered to improve O_2 carrying capacity depending on etiology (must be warmed for pts with cold agglutinin disease).
- Normothermia should be strictly maintained in any pt requiring transfusion(s).
- Possible plasmapheresis for acute removal of IgM antibodies in pts with uncontrolled cold agglutinin disease.

Monitoring

- Standard monitors and urine output, CV status, O_2 saturation (pulse oximetry), and temp regulation (avoiding hypothermia)

General Anesthesia

- Choice of anesthetic technique can vary, but all approaches should have the goal of avoiding hypoxia, hypercarbia, acidosis, stasis, low-flow conditions, and hypothermia.
- Avoidance of hypoventilation.

Regional Anesthesia

- Goals for regional anesthesia are the same as for general anesthesia. No specific contraindications.

Postoperative Period

- Supplemental O_2 therapy
- Adequate hydration
- Early ambulation
- Continued temperature regulation
- Active pulm toilet
- Aggressive evaluation and treatment of fever or infection

Anticipated Problems/Concerns

- Acute periop hemolysis; may warrant transfusion.
- Periop sickling event due to hypoxia, acidosis, hypothermia, or low flow. Sickling can be decreased by increasing arterial oxygen tension.
- Hypothermia-induced cold agglutinin hemolysis; decreased by maintaining normothermia.
- Hypoxia and end-organ damage.
- Venous thrombosis, pulm embolism.

Anemia, Megaloblastic

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Risk

- Prevalence: Estimates ranging from 1.7-3.6%.
- Most common cause is vitamin deficiency: 65% vitamin B12; 12% combined folate/vitamin B12; 6% folate.
- Pernicious anemia is less common: 1 in 7500 people in USA develops pernicious anemia each year.
- Prevalence increases with advanced age and in countries with higher rates of malnutrition.

Perioperative Risks

- Risk of severe anemia and coagulopathy.
- Risk of coronary, cerebral ischemia secondary to severe anemia.
- Increased plasma volume as compensatory mechanism can predispose pts to CHF.

Worry About

- Exaggerated effect of myocardial depression from anesthesia.
- Preoperative treatment should include supplementation of B12 and folate or transfusion in setting of severe anemia and emergent surgery.
- Decreased platelet count and coagulopathy.
- Anemia causing MI, stroke, or resp failure.

Overview

- An anemia caused by a failure of DNA synthesis which results in large, structurally abnormal and immature red blood cells called megaloblasts (MCV >100 fL/cell).
- Often WBC and platelet counts are also decreased.