

Purpura, Immune Thrombocytopenic

Evan G. Pivalizza | Olga Pawelek

Risk

- Rare (100:1 million)
- Children: Male > female
- Adults: Female > male (2–4:1).
- Pregnancy: 1:1000 deliveries; 5% of thrombocytopenia in pregnancy, especially if present in first trimester.

Perioperative Risks

- Hemorrhage (case reports put mortality for splenectomy at 1%, one-third of which is related to bleeding).
- Infection and thrombocytosis post-splenectomy
- Retrospective data from Taiwan point to a higher risk of postop mortality (OR 1.89), complications (1.47), increased length of hospital stay (1.73), and ICU admission (1.89). Preop blood/platelet transfusions are associated with increased risk.

Worry About

- Preop corticosteroids, immunosuppressive agents
- Splenectomy
- Hemorrhage (mucosal when platelet count is $<20,000 \times 10^3/\text{mm}^3$; severe risk [intracranial hemorrhage] with platelet count $<10,000 \times 10^3/\text{mm}^3$; suggestion that mortality is increased if platelet count is $<30,000 \times 10^3/\text{mm}^3$).

Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Airway manipulation—potential hemorrhage	Oral bleeding		
CV	Vascular access			
HEME	Thrombocytopenia	Hemorrhage	Petechiae	Platelets $<20\text{--}50,000 \times 10^3/\text{mm}^3$, megakaryocytes, antiplatelet antibody
CNS	Hemorrhage in acute disease, trauma			Imaging if indicated
OB	Controversy predicting neonate at risk (10–15%) and mode of delivery			

Key References: Choi S, Brull R: Neuraxial techniques in obstetric and non-obstetric patients with common bleeding disorders. *Anesth Analg* 109(2):648–660, 2009; Kühne T, Imbach P: Eltrombopag: an update on the novel, non-peptide thrombopoietin receptor agonist for the treatment of immune thrombocytopenia. *Ann Hematol* 89(Suppl 1):67–74, 2010.

Perioperative Implications

Preoperative Preparation

- Consult with hematology. Acute ITP: Steroids, IV IgG ± anti-D to raise platelet count.
- Steroid supplement if already receiving steroids.
- Premedication: Avoid IM injections.
- Pneumococcal, meningococcal vaccine (plus *Haemophilus influenzae* in children) if for splenectomy.

Monitoring

- Routine.
- Protect pressure points and mucosal surfaces.

Airway

- Avoid nasal ET intubation; caution DLT (case report).

Overview

- Acute, intermittent, or chronic (12-mo) immune-mediated thrombocytopenia (accelerated destruction with appropriate megakaryocyte response). Recent appreciation of impaired plt production, leading to treatment to stimulate platelet growth. Dermal, mucosal, and CNS hemorrhage is most critical.
- Obstetric implications include risk of transient neonatal thrombocytopenia.

Etiology

- Antiplatelet IgG autoantibodies target mature platelets and megakaryocytes, leading to premature removal by spleen and RES. TMO produced in the liver as the principal regulator of megakaryocyte development and platelet production, is suboptimal in ITP pts.

Usual Treatment

- Decrease platelet destruction, although risks of immunosuppression and splenectomy are concerning.
- Corticosteroids: Begun at 1 mg/kg per d (30–60% response rate, up to 80% initially).
- IV immunoglobulin G (0.4–1 g/kg per d).
- Anti-D (if pt Rh-positive) is cheaper and easier than IV IgG.

- Splenectomy: Defer as long as possible in children. Now done laparoscopically (requires disruption of spleen into bag before extraction to prevent splenosis). In chronic disease, splenectomy is indicated if steroids cannot be tapered or response to therapy is poor. In acute disease, indicated for failed medical response and platelet transfusion.
- Second line: Rituximab, monoclonal anti-CD20 antibody, increasingly used to treat refractory ITP. Use is associated with infusion-related side effect, risk of progressive multifocal leukoencephalopathy and infections.
- New stimulatory agents mimic effect of TPO.
- Eltrombopag, a TPO receptor agonist approved (by the FDA) for adults (2008) and children >1 y of age (2015) with chronic ITP. Signal transduction different site than TMO receptor.
- Romiplostim, a TPO mimetic, binds directly to the receptor in the same manner as endogenous TPO. FDA-approved in 2008 (risk evaluation and mitigation strategies required).
- Platelets: Very short survival, may temporarily elevate pH count if there is a bleeding emergency.

Adjuvants

- Individual analysis of risk/benefit for neuraxial technique, especially in parturient pts. Authors recommend platelet count range $>50\text{--}100 \times 10^3/\text{mm}^3$ (European recommendation $>80,000 \times 10^3/\text{mm}^3$). Reports of point-of-care testing (e.g., thromboelastography) in decision making.

Postoperative Period

- Risks of thrombocytosis not as crucial as TTP

Anticipated Problems/Concerns

- Massive surgical hemorrhage
- CNS and airway hemorrhage

Purpura, Thrombotic Thrombocytopenic

Evan G. Pivalizza | Omonle O. Nwokolo

Risk

- TTP rare (1:1,000,000). Adult, pregnancy may be a predisposing factor. Survival 80% at 6 mo; 90% mortality if untreated.
- Periop risks
- Rarely reported in pregnancy or postsurgically (case reports point to cardiac/urologic complications). Refer pt for splenectomy if medical therapy fails. Risks include MAHA with variable neurologic deficits and renal dysfunction combined with thrombocytopenia.

Worry About

- Preop drugs, therapies (plasma exchange, steroids, rituximab)
- CNS/renal dysfunction
- Thrombocytopenia (although usual quantitative platelet transfusion triggers do not apply)

Overview

- Severe MAHA characterized by thrombocytopenia, variable multisystem organ involvement (particularly

CNS and kidney); may be considered part of the spectrum of HUS. Differential in pregnancy from HELLP.

Etiology

- Low ADAMTS13 activity ($<10\%$), a metalloprotease that cleaves multimers of von Willebrand factor, thus leading to increased intravascular platelet aggregation in high-shear environments.
- May be associated with ticlopidine or malignancy.
- Has been described after cardiac, vascular, and abdominal surgery.