Immune Thrombocytopenia

ITP · A diagnosis of exclusion — and treat the bleeding, not the number

TREAT BELOW 30
$\times 10^9$/L, or if bleeding

EXCLUDE EVERYTHING ELSE FIRST *there is no confirmatory test for ITP*

STEP ZERO — IS IT REAL?

Exclude pseudothrombocytopenia before anything else. EDTA-dependent platelet clumping gives a falsely low automated count. **Look at the film for clumps**, and if present **repeat in citrate or heparin**. A manual count on the same clumped sample is **also** falsely low — the fix is removing the clumping, not changing the counting method.

THE ITP PICTURE

Isolated thrombocytopenia with an otherwise **normal blood count and normal film**. No anemia, no leukopenia, no blasts, **no schistocytes**. Spleen not enlarged. Bleeding is **mucocutaneous** — petechiae, purpura, epistaxis, menorrhagia — not deep or joint bleeding.

WHAT TO SEND

Blood film (essential), **HIV, hepatitis C and hepatitis B**, ***H. pylori*** where prevalent, immunoglobulins, liver function, thyroid function, and a pregnancy test. **Bone marrow examination is not routinely required** in a typical presentation.

THE MIMICS *each is managed completely differently*

Consider	Distinguishing feature
Drug-induced	**Heparin (HIT), quinine, sulfonamides, vancomycin, linezolid, valproate.** Review every drug started in the preceding 2 weeks — **stopping the drug is the treatment**.
TTP or other microangiopathy	**Schistocytes with anemia and a raised LDH.** A completely different emergency requiring plasma exchange — **and platelet transfusion is contraindicated**. Look at the film.
DIC	**Abnormal PT, aPTT and fibrinogen with a raised D-dimer.** ITP has a normal coagulation screen.
Liver disease and hypersplenism	Splenomegaly, stigmata of chronic liver disease, other cytopenias. **Thrombopoietin production falls** in cirrhosis.
Marrow disorder	Other cytopenias, blasts or dysplastic cells on the film, constitutional symptoms. **Consider marrow examination in atypical presentations, older patients, or treatment failure.**
Secondary ITP	**Lupus, antiphospholipid syndrome, CLL, lymphoma, common variable immunodeficiency**, and post-viral or post-vaccination. Treat the underlying condition alongside.

TREAT BY BLEEDING AND COUNT *not everyone with a low platelet count needs treatment*

1. **Observation is appropriate for asymptomatic patients with a platelet count above 30 $\times 10^9$/L.** Many need no treatment at all, and the toxicity of therapy often exceeds the risk of the count.
2. **Treat when the count is below 30 $\times 10^9$/L**, or at any count with **clinically significant bleeding** or an upcoming procedure.
3. **First line is corticosteroids** in newly diagnosed adults with a count under 30 who are asymptomatic or have minor mucocutaneous bleeding. **Add IVIG to steroids when a more rapid rise is required**, and use **IVIG or anti-D as first-line if steroids are contraindicated**.
4. **Second line, for disease lasting 3 months or longer:** a **thrombopoietin receptor agonist is preferred over rituximab**, and **rituximab is preferred over splenectomy**. Splenectomy has moved down the sequence, and is usually deferred at least 12 months given the chance of spontaneous remission.
5. **Life-threatening bleeding:** give everything together — **platelet transfusion plus IVIG plus high-dose intravenous corticosteroid**. Platelets are consumed rapidly, so this is the one setting where transfusing them is right, and it works better combined with IVIG. Add tranexamic acid for mucosal bleeding, and consider a TPO agonist early.

PRACTICAL POINTS *the parts that affect day-to-day care*

BEFORE SPLENECTOMY OR RITUXIMAB

Vaccinate against encapsulated organisms — pneumococcus, meningococcus and Haemophilus — ideally at least 2 weeks before splenectomy, and provide **lifelong post-splenectomy advice** with antibiotic prophylaxis. Before rituximab, screen for **hepatitis B** to avoid reactivation, and check immunoglobulins.

ADVICE AND MONITORING

Avoid antiplatelets and NSAIDs. Discuss contact sport and injury risk, and manage menorrhagia actively. **Steroids should not be a long-term maintenance strategy** — taper deliberately and move to a steroid-sparing option rather than settling on a maintenance dose. In pregnancy, involve hematology and obstetrics jointly.

PROCEDURES & PREGNANCY *the two questions you will be asked*

Situation	Approach
Target counts for procedures	Broadly: **> 50 $\times 10^9$/L** for most surgery and invasive procedures, **> 80–100** for neurosurgery and neuraxial anesthesia, and **> 20–30** for minor procedures such as dental extraction or central line insertion. **Plan ahead** — raise the count with steroids or IVIG before the procedure rather than transfusing on the day.
Pregnancy	**Distinguish ITP from gestational thrombocytopenia**, which is far commoner, usually mild (> 70–80), appears in the third trimester and needs no treatment. ITP can occur at any stage and can be more profound. **Corticosteroids and IVIG are the mainstays**; **avoid rituximab and TPO agonists** unless specialist-directed. Manage jointly with obstetrics and hematology, and note that **the neonate can be thrombocytopenic** from transplacental antibody.
Steroid-related harm	Prolonged corticosteroids cause **hyperglycemia, weight gain, mood disturbance, osteoporosis, infection and adrenal suppression**. **Give bone and gastric protection** and plan the exit strategy at the point you start, not months later.

PEARLS & PITFALLS

- **Look at the film before treating.** Clumps mean pseudothrombocytopenia; schistocytes mean you are about to treat the wrong disease.
- **Do not transfuse platelets routinely.** They are consumed within hours and are reserved for serious bleeding — and are actively harmful in TTP and HIT.
- **Treat the patient, not the number.** An asymptomatic count above 30 usually needs observation, not steroids.
- **Review the drug chart.** Drug-induced thrombocytopenia is common, and stopping the drug is curative.
- **TPO agonists now precede rituximab, and rituximab precedes splenectomy** — the second-line order changed.
- **Vaccinate before splenectomy**, and screen for hepatitis B before rituximab.

References: American Society of Hematology 2019 guidelines for immune thrombocytopenia (Blood Advances, 2019) · International consensus report on the investigation and management of primary immune thrombocytopenia.

Educational aid; verify against local hematology protocol and patient factors.