

PART I

Management & Stabilization

Addressing the therapeutic pathway, emergency stabilization protocols, specific interventions, and blood transfusion management guidelines.

Core Principles of Clinical Management

Maintain Cardiorespiratory Stability

- Prioritize airway, breathing, and circulation (ABC).
- Ensure adequate tissue oxygenation and stabilize hemodynamics immediately in acute or severe distress.

Treat Infections Promptly

- Identify and manage co-existing bacterial, viral, or parasitic infections (e.g., malaria, sepsis) early.
- Prevents ongoing bone marrow suppression and further clinical deterioration.

Avoid Unnecessary Transfusions

- Adhere to restrictive transfusion protocols.
- Reserve red blood cell administration strictly for severe anemia accompanied by hemodynamic compromise or clinical shock.

Baseline Hematology Before Empiric Therapy

- Collect essential blood samples (FBC, reticulocyte count, peripheral blood film) prior to starting empiric iron or vitamin supplementation.
- Prevents masking the underlying diagnosis and ensures appropriate targeted treatment.

Clinical Decision Algorithm

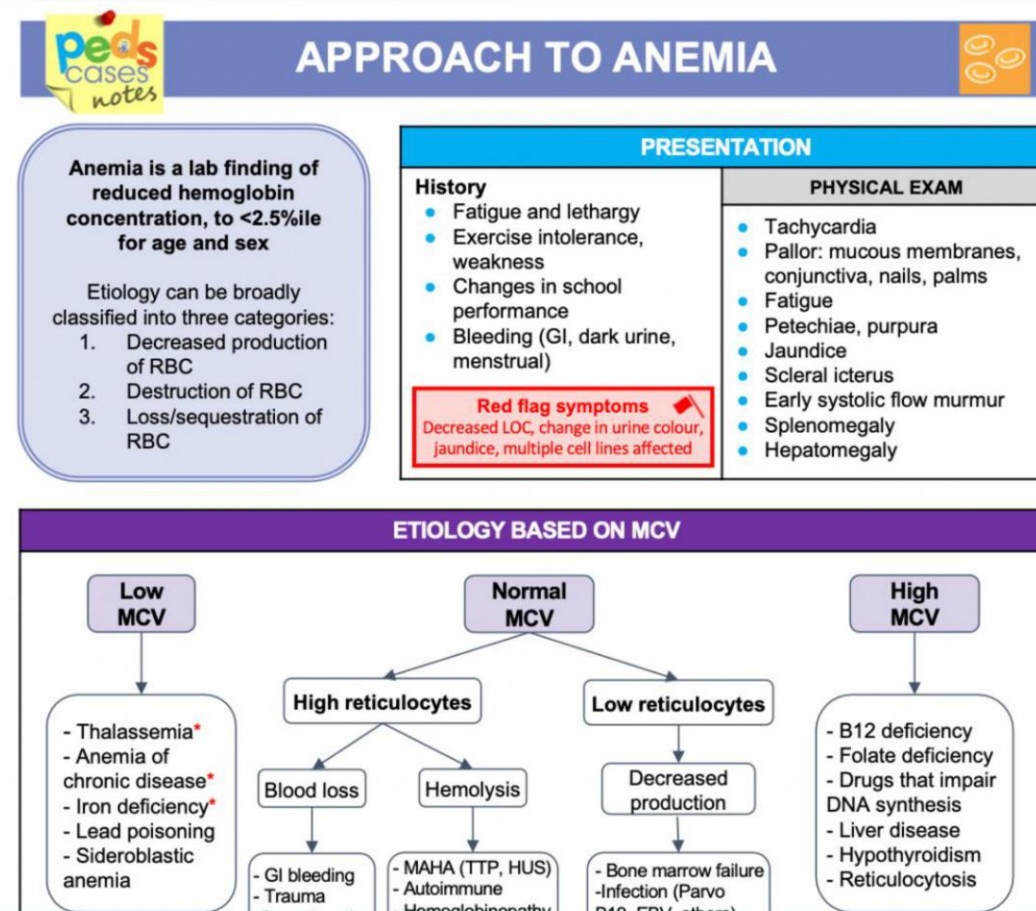
The Reticulocyte Compass

Step 1: Check Reticulocyte Count — Differentiate hypoproliferative bone marrow from hemolytic states or acute bleeding.

Low/Normal Reticulocytes: Suggests nutritional deficiencies (iron, B12, folate), chronic systemic disease, or pure bone marrow failure.

High Reticulocyte Count: Points to peripheral red cell destruction (immune or non-immune hemolysis) or rapid hemorrhage.

Step 2: MCV Categorization — Narrow the marrow failure etiology by reviewing mean corpuscular volume (Microcytic, Normocytic, Macrocytic).



PROFESSOR'S COMMENTARY

"Always utilize the reticulocyte count as your initial compass. If the bone marrow isn't producing reticulocytes in response to low hemoglobin, the system is lacking raw ingredients (iron, B12) or the marrow factory itself is suppressed. Conversely, a high count implies the factory is functional but peripheral destruction is occurring."

- Reticulocytes
- +/- Peripheral blood smear

B) Hemodynamically unstable or severely symptomatic

Cause-Specific: Iron Deficiency Anemia (IDA)

Diet: Limit cow's milk to 500–700 mL/day in toddlers. Encourage iron-rich foods and vitamin C to enhance iron absorption.

Oral iron: Elemental iron 3–6 mg/kg/day in 1–2 doses. Give between meals when possible. - ***Duration:*** *Continue iron for 2–3 months after haemoglobin normalizes to replenish iron stores.*

IV iron: Reserved for children with malabsorption, intolerance or failure of oral iron, inflammatory bowel disease, or chronic kidney disease receiving erythropoietin

Cause-Specific: Iron Deficiency Anemia (IDA)

Vitamin B12 Therapy: Administer intramuscular cyanocobalamin (100–1000 mcg) if malabsorption is suspected.

Folic Acid Support: Oral folic acid 1–5 mg daily is highly effective for systemic deficiency.

Response Monitoring: Expect a prominent reticulocyte spike within 72 to 96 hours post-initiation of therapy.

▮▮ Cause-Specific: Hemolytic & Bone Marrow

Folic Acid Supplementation: Prophylactic daily folic acid for chronic hemolytic anemias (e.g., sickle cell disease, hereditary spherocytosis) due to high cellular turnover.

Specialized Care: Multidisciplinary management including hematology consultation, vaccination against encapsulated organisms (pneumococcus, meningococcus, Hib) for asplenic children, and specialized crisis management (hydroxyurea, exchange transfusions for severe sickle cell complications).

🔪 Cause-Specific: Hemolytic & Bone Marrow

Hemolytic States (G6PD & Sickle Cell)

Oxidative Trigger Avoidance: For G6PD, strictly avoid fava beans, naphthalene, sulfa drugs, and anti-malarials.

Vaso-occlusive Prophylaxis: Hydroxyurea to boost Hemoglobin F, aggressive hydration, and folic acid supplementation.

Infection Control: Oral penicillin prophylaxis and urgent evaluation of febrile episodes.

Autoimmune & Marrow Failures

Immune Cytopenias (AIHA): Initiate systemic corticosteroids (Prednisolone 2 mg/kg/day) to blunt antibody production.

Marrow Suppression: Supportive care, isolation protocols to prevent infections, and prompt bone marrow biopsy.

Malignancy Workup: Urgent pediatric hematology referral if blast cells are present on the blood smear.

PROFESSOR'S COMMENTARY

"In G6PD deficiency, the clinical course is usually self-limiting as older red blood cells lyse first. However, watch out for acute intravascular hemolytic crises which can result in hemoglobinuria (Coca-Cola urine) and acute kidney injury due to free hemoglobin blocking the renal tubules."

Blood Transfusion Hemoglobin Thresholds

Severe Chronic Anemia (Asymptomatic)	Hb < 5.0 g/dL
Acute Symptomatic Anemia (Symptom-driven)	Hb < 7.0 g/dL
Severe Pulmonary / Cyanotic Heart Disease	Hb < 9.0 g/dL

Blood transfusion is not a treatment for nutritional anemia unless specific emergency criteria are met

PROFESSOR'S COMMENTARY

"Transfusion is a clinical decision, never a purely numeric one. A child with severe chronic iron deficiency with a hemoglobin of 4 g/dL can play comfortably because their heart has had months to adjust. Infusing blood rapidly into this child can result in high-output cardiac failure due to sudden volume overload."

Indications for Packed Red Blood Cell (PRBC) Transfusion

➤ **Blood transfusion is not a treatment for nutritional anemia unless specific emergency criteria are met**

1. Severe symptomatic anemia with signs of impending cardiovascular collapse
2. Acute, massive blood loss with ongoing hypovolemic instability
3. Pre-operative preparation for children with severe anemia

pRBC Transfusion Dosing & Safety

Standard Dosing & Administration

Transfusion Dosing: Administer **10–15 mL/kg** of packed red blood cells (pRBCs).

Anticipated Response: Expect each 10 mL/kg aliquot to increase hemoglobin by approximately 2 g/dL.

Administration Rate: Infuse slowly over 2–4 hours; never exceed 4 hours to avoid bacterial growth risk.

Critical Safety Guidelines

Severe Chronic Compensated Cases: Reduce volume to **5 mL/kg** and infuse extremely slowly over 4 hours.

Diuretic Administration: Consider co-administering intravenous Furosemide (0.5–1 mg/kg) to mitigate TACO risk.

Vitals Monitoring: Continuously record vital signs before, during, and up to 4 hours post-transfusion.

PROFESSOR'S COMMENTARY

"In chronic severe anemia, the cardiac muscle is chronically hypoxic and extremely sensitive. If you transfuse 15 mL/kg at standard rates, you will trigger pulmonary edema. In these high-risk scenarios, we split the volume into small, slow aliquots, with a dose of furosemide in between."

⚠ Monitoring & Managing Complications

Reaction Type	Clinical Presentation	Pathophysiology	Immediate Intervention
TACO (Circulatory Overload)	Acute respiratory distress, hypoxemia, tachycardia, jugular venous distention	Volume overload leading to acute left ventricular failure	Stop transfusion , place upright, administer high-flow O2 and IV diuretics
Acute Hemolytic	Fever, chills, hemoglobinuria, flank pain, cardiorespiratory collapse	ABO incompatibility leading to massive complement activation	Stop transfusion , run normal saline, support blood pressure, preserve urine output
Febrile Non-Hemolytic	Temperature rise $\geq 1^{\circ}\text{C}$ within 4 hours, chills, mild tachycardia	Cytokines accumulated in stored donor blood	Pause transfusion, administer antipyretics (Acetaminophen), rule out hemolytic cause

PROFESSOR'S COMMENTARY

"If a reaction occurs, the first action is identical across all categories: Stop the transfusion immediately and replace the line with normal saline. Never discard the blood bag or tubing; they must return to the blood bank for compatibility testing."

Emergency Management of Anemic Shock

- 1. Oxygenation Optimization:** Administer 100% high-flow oxygen via non-rebreather mask to saturate remaining hemoglobin.
- 2. Establish Vascular Access:** Secure two large-bore IVs. Initiate Intraosseous (IO) access if peripheral access fails within 90 seconds.
- 3. Cautious Fluid Boluses:** Infuse crystalloids carefully (**10 mL/kg** instead of standard 20 mL/kg) to prevent heart failure.
- 4. Emergency O-Negative Blood:** Give uncrossmatched O-negative pRBCs immediately if bleeding is catastrophic.



Signs of Decompensated Shock

Monitor closely for signs of organ hypoperfusion:

- Lethargy, irritability, or obtundation
- Severe tachycardia progressing to bradycardia
- Cold extremities & capillary refill > 3 seconds

PROFESSOR'S COMMENTARY

"When resuscitating a child in shock secondary to chronic anemia, remember the heart muscle is already hypoxic. Flooding the system with standard 20 mL/kg crystalloid boluses can dilate the ventricles and trigger arrest. We use smaller boluses of 10 mL/kg and monitor the lungs closely for crackles."