



Haematological Disorders of Childhood

Anaemia

WHO criteria: Hb < 11g/dL

Iron def anaemia (poor diet, poverty, bleeding)

Thalassaemia (Mediterranean, SE Asian, hepatosplenomegaly)

MCV > 70: Microcytic

Haemolysis

Marrow failure (following infection, malignancy, thyroid / kidney / liver failure)

MCV 81 - 97: Normocytic

Folate def (malabsorption)

Signs of B12 def: poor feeding, late milestones, odd movements, microcephaly, failure to thrive

B12 def (breast milk from vegie, low intrinsic factor, malabsorption)

MCV > 100: Macrocytic

CNS iron levels fall before RBC mass

Check ferritin

Even if a child is not anaemic they still may be iron deficient

Treating low ferritin may improve: memory, developmental delay, mood, cognition

Haemolytic Anaemia

Malaria

Main cause of haemolysis in N Europe

Autosomal dominant

Splenectomy indicated in severe disease and leads to increased RBC survival

Hereditary Spherocytosis

Homozygote have sickle cell anaemia

Heterozygote have sickle cell trait

Autosomal recessive

Common in people of African origin

If carrier protect from falciparum malaria and causes no disability except in hypoxia (unpressurised aircraft / anaesthesia) where vaso occlusive events may occur (check all of African descent pre op)

Sickle Cell

Pathogenesis: deformed RBCs, fragile and haemolyse, block small vessels

Signs and symptoms are highly variable

Vaso-occlusive painful crisis: common due to microvascular occlusion, often affects bone marrow causing severe pain

Complications: splenic infarction (and increased susceptibility to infection), growth impairment, bone necrosis, chronic renal failure, long term lung damage

ALL

Commonest childhood leukaemia

Pancytopenia (pallor, infection, bleeding), fatigue, anorexia, fever, bone pain

Presentation

Period prior to diagnosis is often brief (2 - 4 weeks)

Derranged WCC (although can be normal), normochromic normocytic anaemia, low platelets

Tests

Marrow: 50 - 98% nucleated cells will be blasts; CSF: pleocytosis

Risk assessment to ensure only those likely to relapse get the more aggressive four drug option +/- stem cell transplant

Remission induction: 4 weeks of chemo

Consolidation phase: cranial irradiation is known CNS disease + further chemo

Continuation phase: monthly chemo and short courses of oral steroids for 2 1/2 years

Treatment

Complications

Infection: neutropenic regimen, co-trimoxazole prevents pneumocytosis, revaccinate 6 months after chemo

Hyperuricaemia from massive cell death on induction

Poor growth

Cancer elsewhere

Relapse

Prognosis

Worse prognosis

Black, extremes of age, male, WCC > 100, philadelphia translocation

80% cure