Haematological Disorders of Childhood

Anaemia

- WHO criteria: Hb < 11g/dL
- MCV > 70: Microcytic
  - Iron def anaemia (poor diet, poverty, bleeding)
  - Thalassaemia (Mediterranean, SE Asian, hepatosplenomegaly)
- MCV 81 - 97: Normocytic
  - Haemolysis
  - Marrow failure (following infection, malignancy, thyroid / kidney / liver failure)
- MCV > 100: Macrocytic
  - Folate def (malabsorption)
  - B12 def (breast milk from vegie, low intrinsic factor, malabsorption)
  - Signs of B12 def: poor feeding, late milestones, odd movements, microephaly, failure to thrive
- Even if a child is not anaemic they still may be iron deficient
  - CNS iron levels fall before RBC mass
  - Check ferritin
  - Treating low ferritin may improve: memory, developmental delay, mood, cognition

Haemolytic Anaemia

- Malaria
- Hereditary Spherocytosis
  - Main cause of haemolysis in N Europe
  - Autosomal dominant
  - Splenectomy indicated in severe disease and leads to increased RBC survival
- Sickle Cell
  - Autosomal recessive
    - Homozygote have sickle cell anaemia
    - Heterozygote have sickle cell trait
  - 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)
  - 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
- Presentation
  - Pancytopenia (pallor, infection, bleeding), fatigue, anorexia, fever, bone pain
  - Period prior to diagnosis is often brief (2 - 4 weeks)
- Tests
  - Derranged WCC (although can be normal), normochromic normocytic anaemia, low platelets
  - Marrow: 50 - 98% nucleated cells will be blasts; CSF: pleocytosis
- Treatment
  - 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
- Complications
  - Infection: neutropenic regimen, co-trimoxozole 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