

Anaemia / SCD/ Bleeding disorders in Children

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Learning Outcomes

- Describe mechanisms of anaemia in Malawian children
 - Discuss important causes of anaemia
 - Describe presentation and Rx of SCD and G6PD in Malawi
 - Identify causes of anaemia from history and FBC/Film
 - Discuss common causes of bleeding diathesis in children
 - Prescribe blood for severe anaemia according to WHO guidelines
- HANDOUT**

Mechanisms of Anaemia?

- Blood Loss
- Haemolysis
- Reduced red cell production:
 - Nutrient deficiency
 - Marrow suppression / failure

Causes of Anaemia

Blood Loss

- Hookworm
- Schisto
- epistaxis
- Gastritis / Meckels / colitis

Haemolysis / consumption

- Malaria
- Defective red cell:
 - Membrane: Spherocytosis
 - Enzymes: G6PD
 - Haemoglobin: SCD, Thalassaemia
- Hypersplenism

Reduced Red cell production

- Nutrient deficiency
 - Iron
 - B12 / Folate
 - PEM
- Marrow suppression
 - HIV
 - Sepsis
 - Malaria
 - Miliary TB
 - Renal Failure
 - Leukaemia
 - Parvovirus B19
 - Chloramphenicol etc...

Severe Anaemia Mechanisms in Malawian Children

Red cell production failure 48%:

- Bacteraemia
- HIV
- Malaria
- Wasting
- Vit B12 def
- Vit A def

Haemolysis 21.7%:

- Malaria
- G6PD in boys

Blood Loss 7.2%:

- Hookworm

Calis et al NEJM 2008;358(9):888-9

Severe Anaemia (Hb <5g/dL) Malawian Children

	Patients (%)	Controls (%)
Wasting	16	6
Iron Deficiency	47	69
B12 deficiency	30	16
Folate deficiency	0	0
Vit A deficiency	92	66
HIV	13	6
Bacteraemia	15	4
Malaria	59	43
Hookworm	10	1.9
Schistosomiasis	1.3	1.2
G6PD	14	9

Severe Anaemia in Malawian Children

- Malaria major factor
- Bacteraemia with NTS common
 - Due to dyserythropoeisis
 - Consider antibiotics in febrile anaemic child, even if MPS +ve
- VIT A + B12 deficiency significant problems
- G6PD important cause severe anaemia
- HIV causing severe anaemia associated with high mortality
 - 18 months after presentation with Hb <5g/dL, 65% of the HIV +ve had died, 10.7% of the HIV –ve (pre-ART study)
- Hookworm common in all anaemic > 1 year

What about Iron?

- Fe deficiency causing mild anaemia is common (PCV > 20%) and associated with hookworm
- May contribute to poor school performance
- Fe supplements reduce anaemia prevalence in community
- Fe supplements may increase mortality in areas with high malaria / sepsis burden
- Iron deficiency may protect against severe anaemia, as ?reduces bacteraemia risk
- Iron should *not* be given acutely to children with malaria / NTS sepsis / malnutrition
- *Wait for child to recover – then give for 3 months*

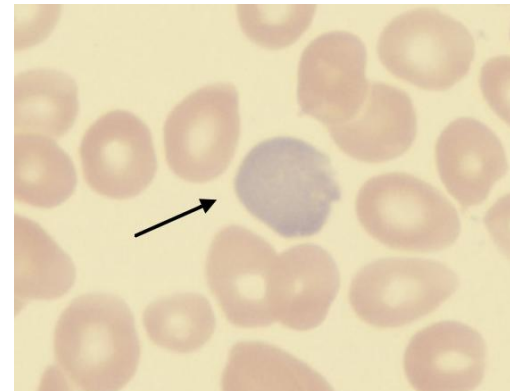
Working out the cause of the anaemia

- Red cell production:
 - Film:
 - Microcytic hypochromic: Fe
 - Macrocytic, hypersegmented neut: B12 / folate
 - Blasts: Leukaemia
 - Malarial parasites
 - Target cells, fragments, reticulocytes
 - FBC:
 - ↓ WBC / platelets: marrow failure
 - ↓ reticulocytes

Working out the cause of the anaemia

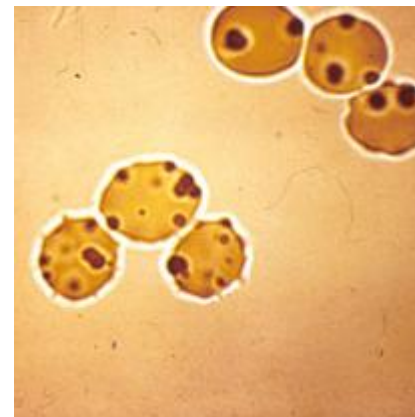
- Haemolysis / consumption
 - Often enlarged spleen
 - Urobilinogen / jaundice
 - Film:
 - Sickle
 - targets (Thall)
 - Malaria
 - Heinz bodies (rare)
 - ↑ reticulocytes
 - Coombs test: autoimmune

- Blood loss:
 - History
 - Stool (HWA)
 - urine (schisto)



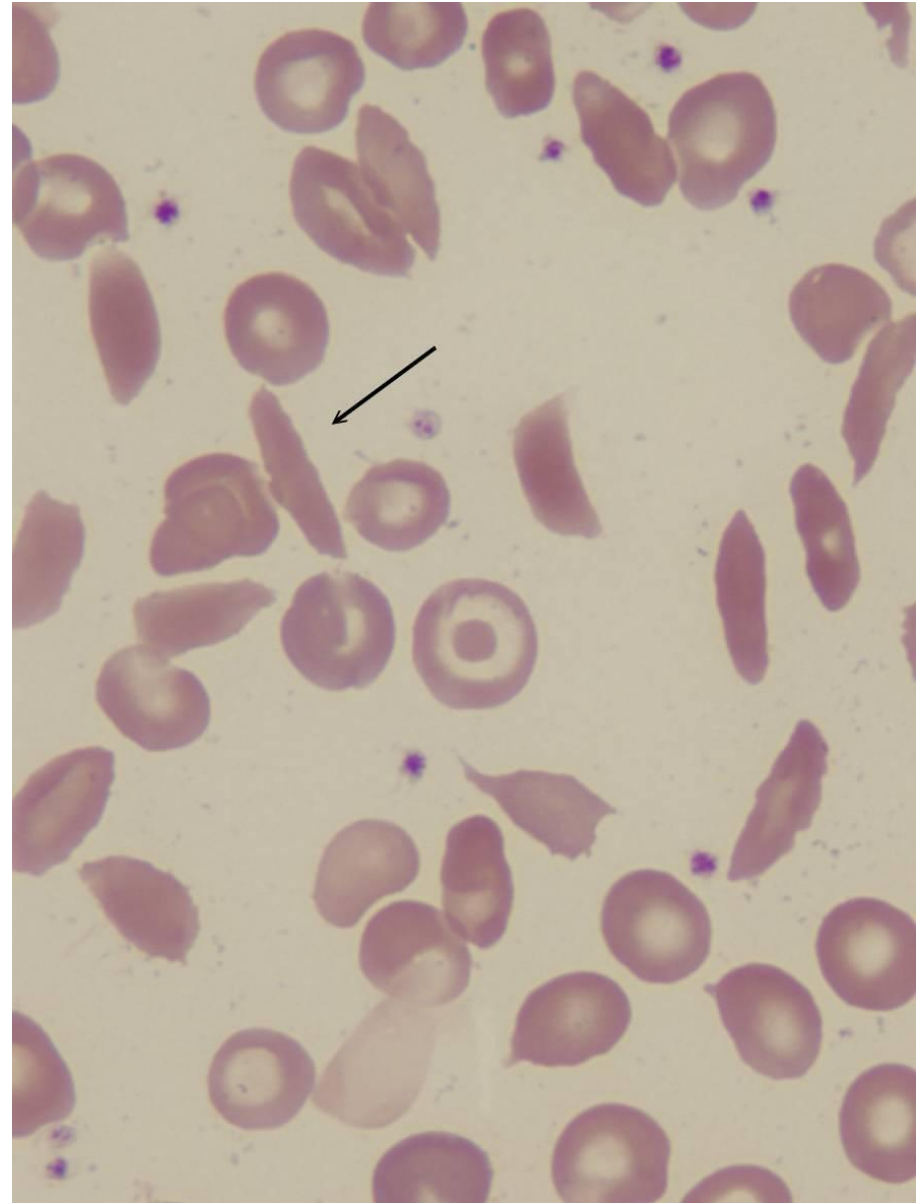
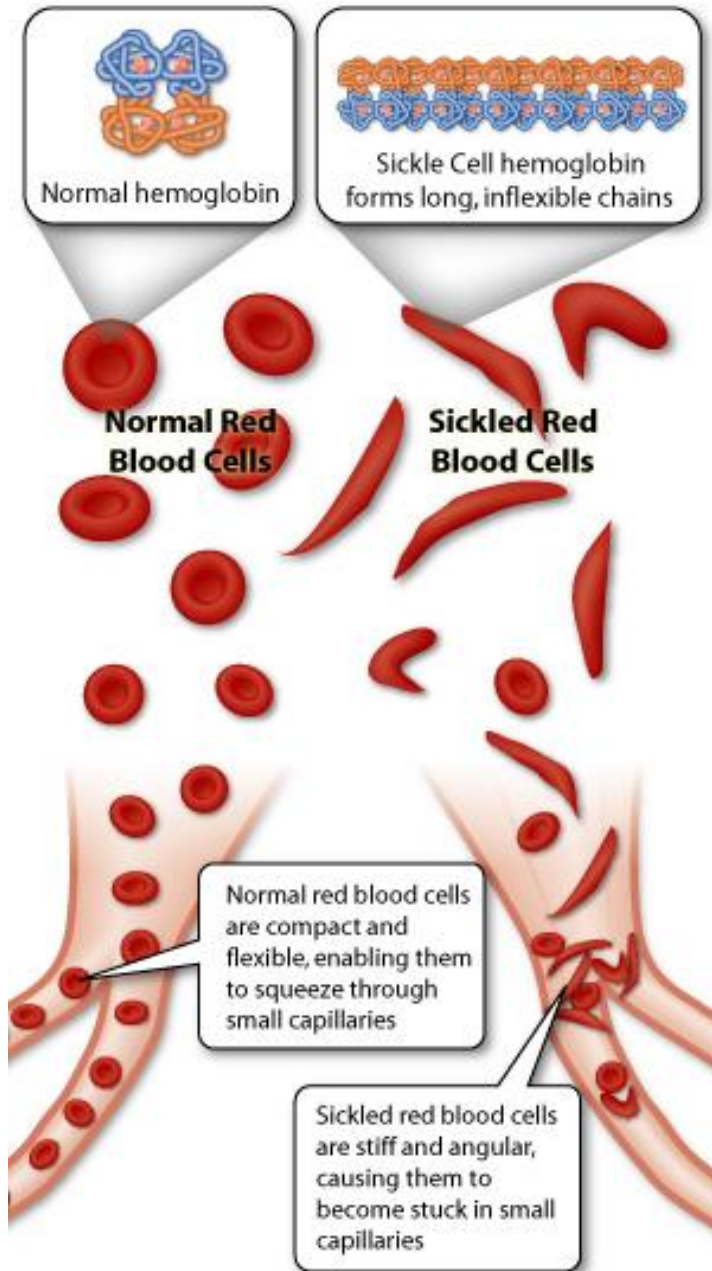
Haemolytic anaemia – G6PD

- Common in Malawi (10%)
- X linked
- Usually mild
- Favism induced by:
 - ASA, SP, CTX
 - Malaria / infections
- Diagnosis:
 - Presumptive
 - Enzyme, Heinz bodies



Haemolytic Anaemia - SCD

- Rarer (1.5%) but more significant
- Autosomal recessive – Hb SS
- Glutamic acid – valine substitution in β chains
- HbS = 2 α chains, 2 defective β chains
- Low oxygen tension leads to:
 - Polymerisation of HbS chains
 - Distortion of red cell shape (sickle)
- Distorted red cells cause ischaemia



Haemolytic Anaemia - SCD

- Symptoms > 6/12
- Common problems:
 - Hand / foot syndrome
 - Anaemia – repeat transfusions
 - Jaundice
 - Infections: Malaria, pneumococcus, NTS
 - Limb / abdo pain
- Less common:
 - Aplastic crisis (parvovirus)
 - Stroke
- Diagnosis



SCD Treatment

- Regular:
 - Benzathine prophylaxis / pneumovacc if avail
 - Malaria prophylaxis (ITN / SP)
 - Folic acid
 - Education on treating GE / RTI
 - Monitor Hb, transfuse needed
 - Hydroxyurea
- Crisis:
 - Analgesia (PCM, ibuprofen, morphine)
 - Fluids – ORS / IV

Case 1

- 18 months old girl, weight 10kg
- Febrile (temp 39.6C) dyspnoeic
- Hb 3g/dl

Case 2

- 15 months old girl
- Pallor
- Hb 4.5g/dl
- MCV 62 fL
- Diet 'normal'

Case 3

- 3 year old boy
- Persistent diarrhoea
- Failure to thrive
- Hepatosplenomegaly
- Axillary lymphadenopathy
- Hb 5g/dl
- MCV 104 fL

Case 4

- 20 months old female child
- Recurrent upper and lower limb pain
- Had one episode of jaundice recently
- Eats poorly
- Generally irritable unhappy and not thriving
- Chronically ill-looking, small for age
- Mild jaundice
- Swollen hands
- HB 6g/dl

Case 5

- 10 year old boy
- General malaise
- Weakness
- Bone pain
- Bruises easily
- Hb 6 g/dl
- WCC 45×10^9 /L
- Platelets 23×10^9 /L

Case 6

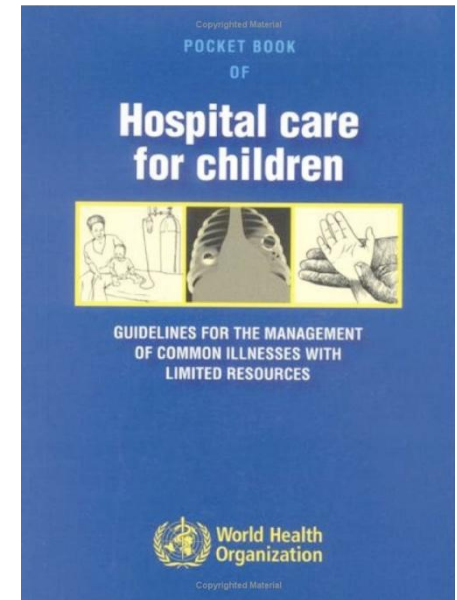
- 4 year old girl
- Generalised lymphadenopathy
- Oropharyngeal candidiasis
- Recurrent epistaxis
- Hb 7.5 g/dl
- WCC 17
- Platelets 29

WHO treatment chronic anaemia

- Fe or FeFol daily x 14 days
- Review at 2/52
- Continue Fe / FeFol x 3 months
- Deworm if > 2years

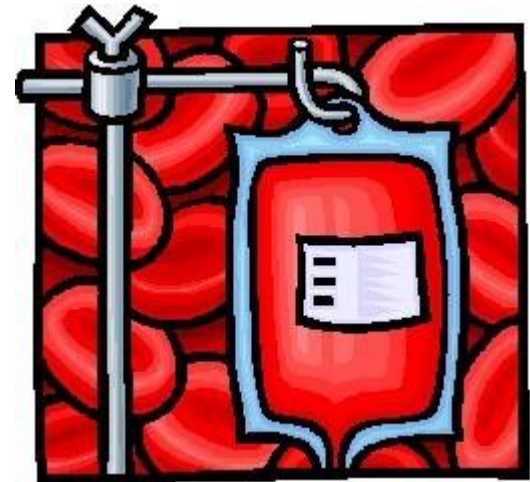
BUT in Malawi.....

- Vit A and B12 deficiency may be more important
- Hookworm common in under 2s



WHO treatment severe anaemia

- Transfuse if:
 - PCV < 12% *or*
 - PCV 13-18% and:
 - Clinically severe dehydration
 - Shock
 - Impaired consciousness
 - CCF
 - Deep / laboured breathing
 - Parasitaemia > 10%



WHO treatment severe anaemia

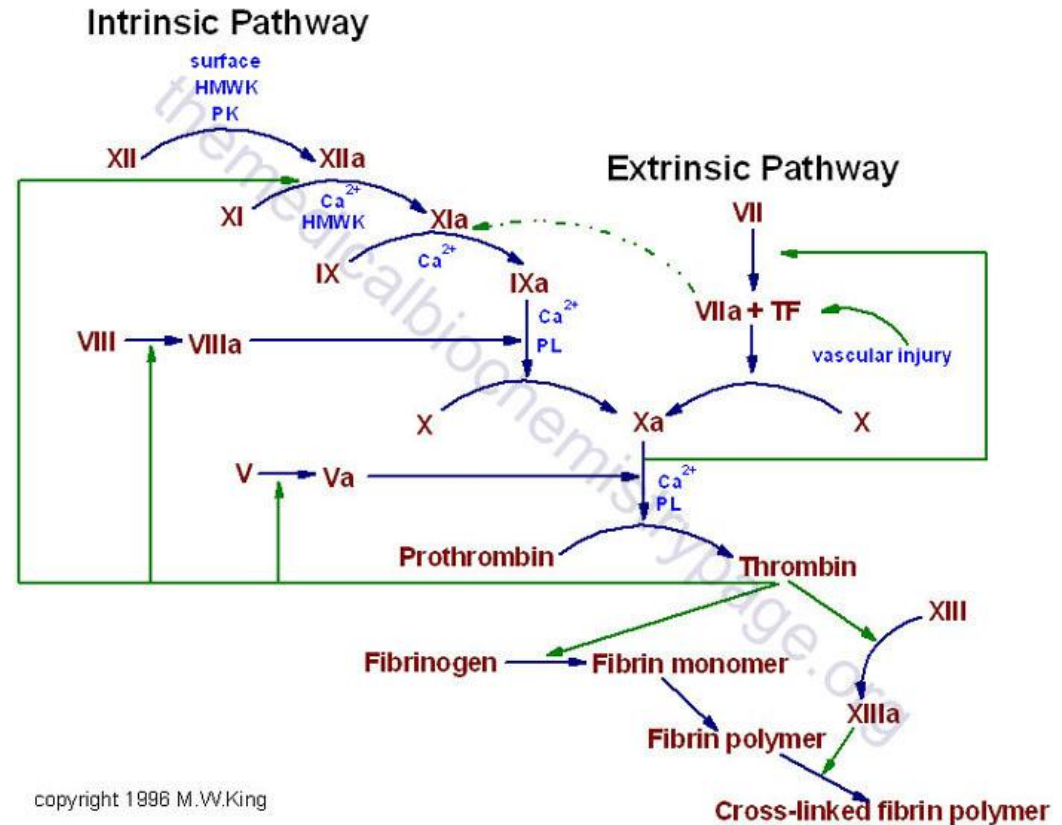
- If NOT malnourished:
 - 10mls/kg packed cells over 3-4 hours *or*
 - 20mls/kg whole blood
 - Frusemide only if signs of overload
- If malnourished:
 - 10 mls/kg whole blood
 - FLUID OVERLOAD COMMON – give Frusemide

Things to think about in a child with 'SMA'

- Is it really *just* malaria?
- Is this the first transfusion?
- Have they got SCD or G6PD?
- Have they got HIV?
- Have they got septicaemia?

Bleeding diathesis

- Coagulation requires:
 - Normal platelets (number and function)
 - Normal clotting factors



Platelet disorders

- Present with mucosal bleeding / purpura
- Causes include:
 - Infection and platelet destruction
 - DIC
 - Marrow infiltration (leukaemia)
 - ITP / HIV autoimmune thrombocytopenia
 - Hypersplenism
 - Abnormal platelet function – von Willebrands

Clotting cascade disorders

- Hereditary – Haemophilia
 - Haem A – Factor VIII – x linked
 - Spontaneous haemarthroses
 - Rx Cryoprecipitate
- Acquired – Vit K deficiency
 1. Decreased absorption fat soluble vits (hepatic failure)
 2. Haemorrhagic disease of the newborn
 - Present with GI bleeds, bruising

Bleeding diathesis – Ix

- FBC, coagulation screen, culture
- PT measures EXTRINSIC (*PET*) – *hepatic failure, warfarin*
- PTTK measures INTRINSIC – *heamophilia (factor 8), heparin*
- DIC – PT, PTTK and platelets affected

Bleeding diathesis – Rx

- Platelet problems:
 - Platelet transfusion if < 20 (not ITP)
 - Consider steroids
 - Immunoglobulin for ITP
- Clotting factor deficiencies:
 - Vit K for hepatic failure, HD Newborn
 - Factor 8 cryo for haemophilia
 - Fresh Frozen Plasma in emergency

Mafumbo ghalipo?



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