Haematology Cases for GPs

Jane Norman

October 2016

- 10 mixed haematology cases
  - Common GP questions
  - Abnormalities to look out for
  - What happens to patient in secondary care
- Useful links and referral process for UHBristol
- Cases or queries

CASE 1

20 year old female, presents with petechial rash

Hb 122 (115-170) MCV 86 (81-98) WCC 5 (4-11) Plts 6 (150)

Normal coagulation screen

Differential diagnosis?

Further tests?

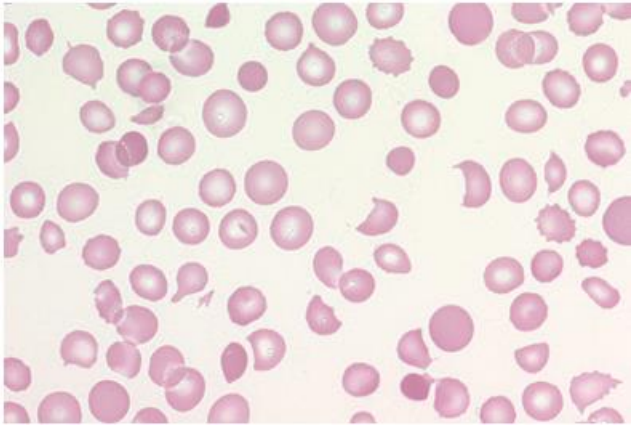
- ITP
  - Idiopathic
  - Associated with lymphoma
- Microangiopathic haemolytic anaemia
  - Thrombotic thrombocytopenic pupura/Haemolytic Ureamic syndrome
- Marrow infiltration
  - Acute leukaemia
- Drugs
- Infection

Immune thrombocytopenia

- Children- spontaneously resolve
- Adults (tx below 30)
  - 80% respond to steroid
  - 30% relapse
  - Tx options include immunosupression, splenectomy, TPO agonists

MAHA

1. Haemolytic anaemia
2. Thrombocytopenia
3. Fever
4. Neurological symptoms
5. Renal impairment



TTP

More significant thrombocytopenia

Urgent plasma exchange

HUS

Diarrhoea (E.coli 0157)

Renal physicians

CASE 2

45 year old man, presents with tiredness

Hb 140 (130-170), WCC 7 (4-11), Plts 358 (150-450)
Ferritin 1052 (30-400)

No PMH

Well

No significant alcohol hx

No known liver disease

Haemochromatosis

- Autosomal recessive
- Mutation on HFE gene (C282Y 90% of cases)
- Heterozygotes not at risk from iron overload, but consider family members

- Homozygotes
  - Persistently raised ferritin in absent of inflammatory causes (liver disease can be tricky)
  - Evidence of end organ damage
    - congestive cardiac failure
    - liver dysfunction
    - diabetes
    - hypogonadism
  - Transferrin saturations (>60%)
  - Check HFE gene
  - Pre-menopausal women are self venesecting
  - We aim to keep ferritin < 100 and transferrin saturation < 50%
  - Avoid alcohol
  - Cut down exogenous iron

CASE 3

62 year old man, presents tired

Hb 91 (130-170) MCV 79 (81-98) WCC 6 (4-11) Plts 491(150-450)

White Caucasian

No GI symptoms/blood loss

No weight loss

Good diet

Ferritin: 72 (30-400), B12: Normal, Folate: Normal

CRP 102 (<10); Zinc protoporphyrin (ZPP) 120 (high); reticulocyte-Hb low.

Iron 3 (8-15); transferrin saturation 10%; total iron binding capacity 80 (normal)

	Iron Overload	Iron Deficiency	Anaemia of Chronic Disease
Serum Iron 10-30 umol/l	High	Low	Normal (low)
Total iron binding capacity (TIBC) 47-70 umol/l	Low	High	Normal (low)
Transferrin saturation 16-50%	High	Low	Normal

Iron deficient

Anti-TTG antibodies negative

Upper and lower GI scopes normal

Hb 93 MCV 79 WCC 5 Plts 475

Start ferrous sulphate
Cannot tolerate, try ferrous fumarate cannot take
Refer for IV iron

Starts to lose weight
Gastro referral capsule endoscopy- GI tumour

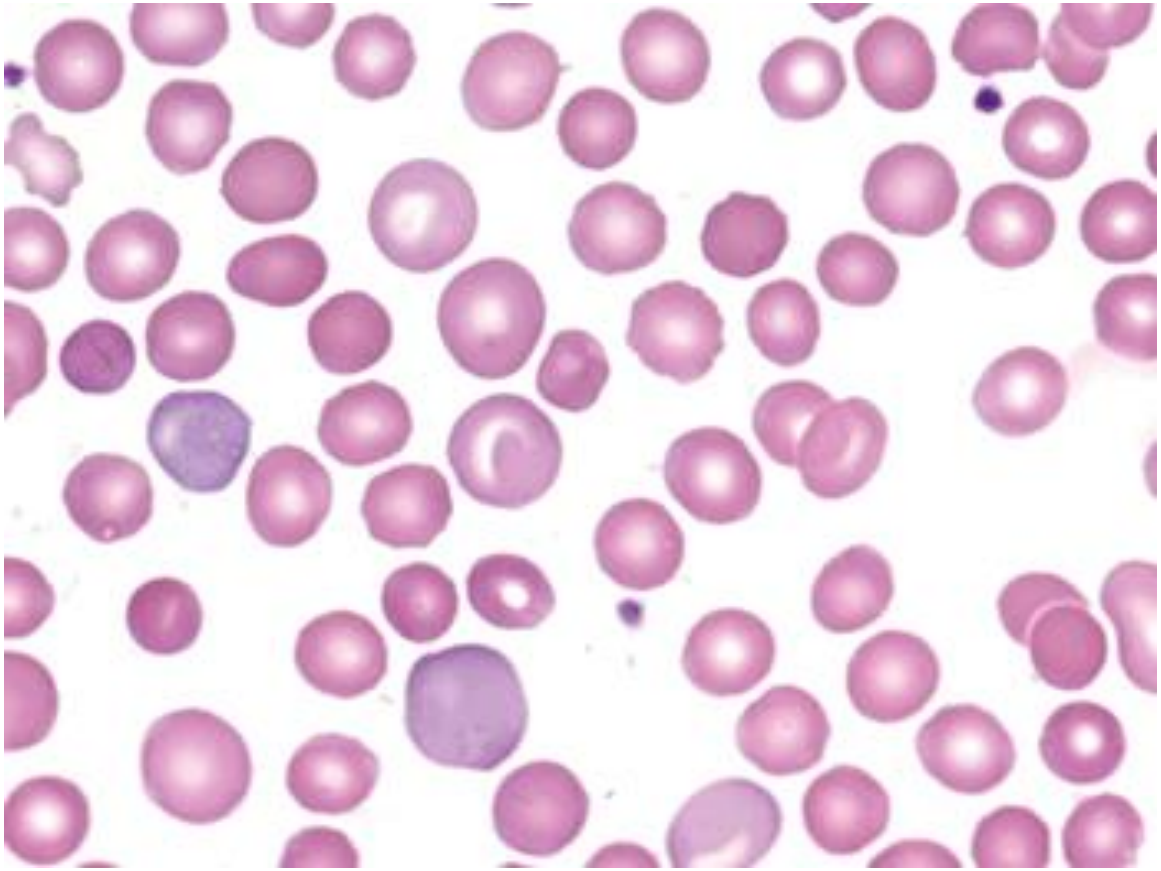
CASE 4

67 yr old white male, presents pale, tired and SOB

Hb 63, MCV 102, WCC 8, Plts 211

Film report:

Polychromasia and spherocytosis, no red cell fragments



Polychromasia and Spherocytes

Haemolysis screen

- FBC/film
- Reticulocyte count
- LDH
- Bilirubin
- Haptoglobin
- Direct antiglobulin test

- Raised
- Raised
- Raised
- Low
- Positive

BRITISH COMMITTEE FOR STANDARDS IN HAEMATOLOGY

BCSH GUIDELINES

Haematology Guidelines

All | Haemato-oncology | General Haematology | Haemostasis and Thrombosis | Transfusion

SHOW 10 ENTRIES | CURRENT GUIDELINES | IN PROGRESS | ALL | SEARCH THIS SECTION

Page : 1 2 3 4 5 6 7 8 9 10 11 12

TOPIC	GUIDELINE TITLE	DATE	FULL GUIDE	PUBLISHED VERSION	AUDIT TEMPLATE
Platelet transfusion	**Use of Platelet Transfusions**	2016			
Transfusion Sickle Cell Disease	**Red Cell Transfusion in Sickle Cell Disease. Part 1: Principles and Laboratory Aspects**	2016			
Transfusion in Sickle Cell Disease	**Red Cell Transfusion in Sickle Cell Disease. Part II: Indications for Transfusion**	2016			
Red Cell Transfusion in Sickle Cell Disease	**Red Cell Transfusion in Sickle Cell Disease. Part 1: Principles and Laboratory Aspects.**	2016			
Red Cell Transfusion in Sickle Cell Disease	**Red Cell Transfusion in Sickle Cell Disease. Part II: Indications for Transfusion**	2016			
Peri-op anti-coagulation and anti-platelet therapy	**Per-Operative Management of Anti-Coagulation and Anti-Platelet Therapy**	2016			
AIHA	**Diagnosis, Investigation and Management of Primary Autoimmune Haemolytic Anaemia**	2016			
AIHA	**Management of drug induced immune and secondary Autoimmune Haemolytic Anaemia**	2016			
Eosinophilia	**Investigation and Management of Eosinophilia**	2016			

HAEMOLYSIS

CONGENITAL

ACQUIRED

NON-IMMUNE

IMMUNE

Immune

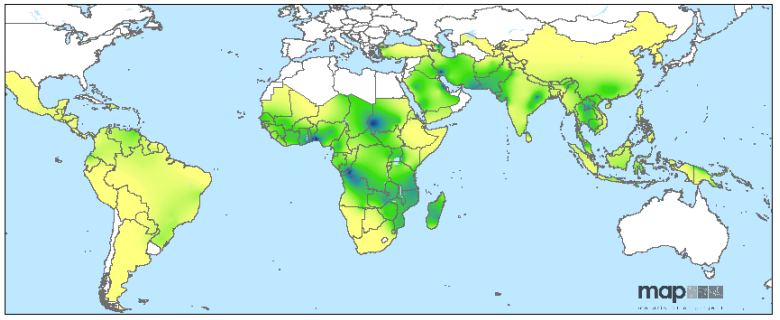
- Autoimmune haemolytic anaemia (Dat +ve)
  - Warm
    - Steroid/immunosuppression
    - Often associated with lymphoproliferative disorder
  - Cold
    - Consider mycoplasma infection
    - Warmed blood samples
- Consider delayed transfusion reaction

Non Immune

- Drugs (dapsone, celphalosporins, levodopa)
- Valve related haemolysis
- March haemolysis
- Microangiopathic haemolytic anaemia
- Paroxysmal nocturnal haemoglobinuria

Congenital

- Haemoglobinopathy
  - Sickle
  - ß-Thalassaemia (intermediate/major)
- Hereditary spherocytosis
  - Northern European
  - Autosomal dominant
  - Gallstones
  - Symptomatic when unwell
- G6PD deficiency
  - West Africa/Middle East/SE Asia
  - X linked recessive
  - Infection/drugs/fava beans



CASE 5

5 yr old girl, known HbSS disease

- Very few preceding sickle complications
- 1 week history of malaise, fevers which have settled
- Now pale, tired, unwell
- Hb 29, WCC 10, Plts 332

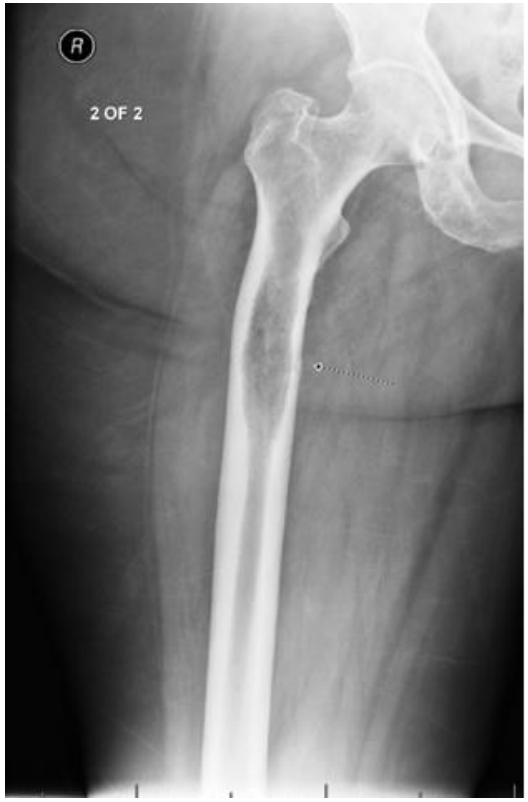
Parvovirus B19

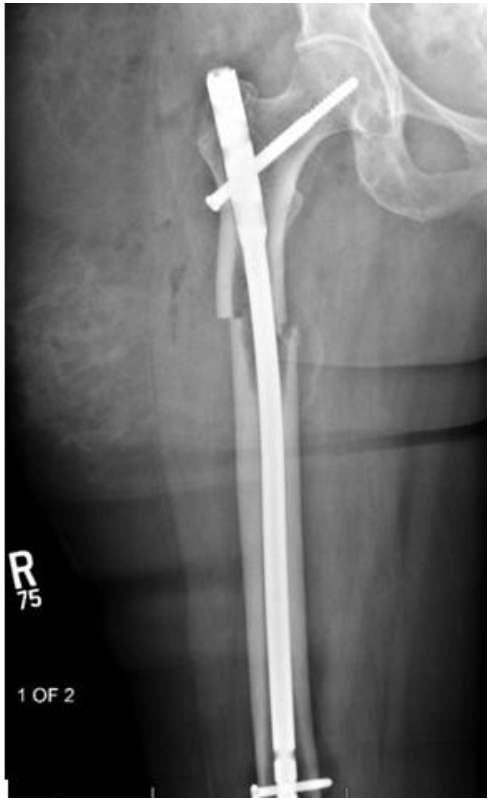
CASE 6

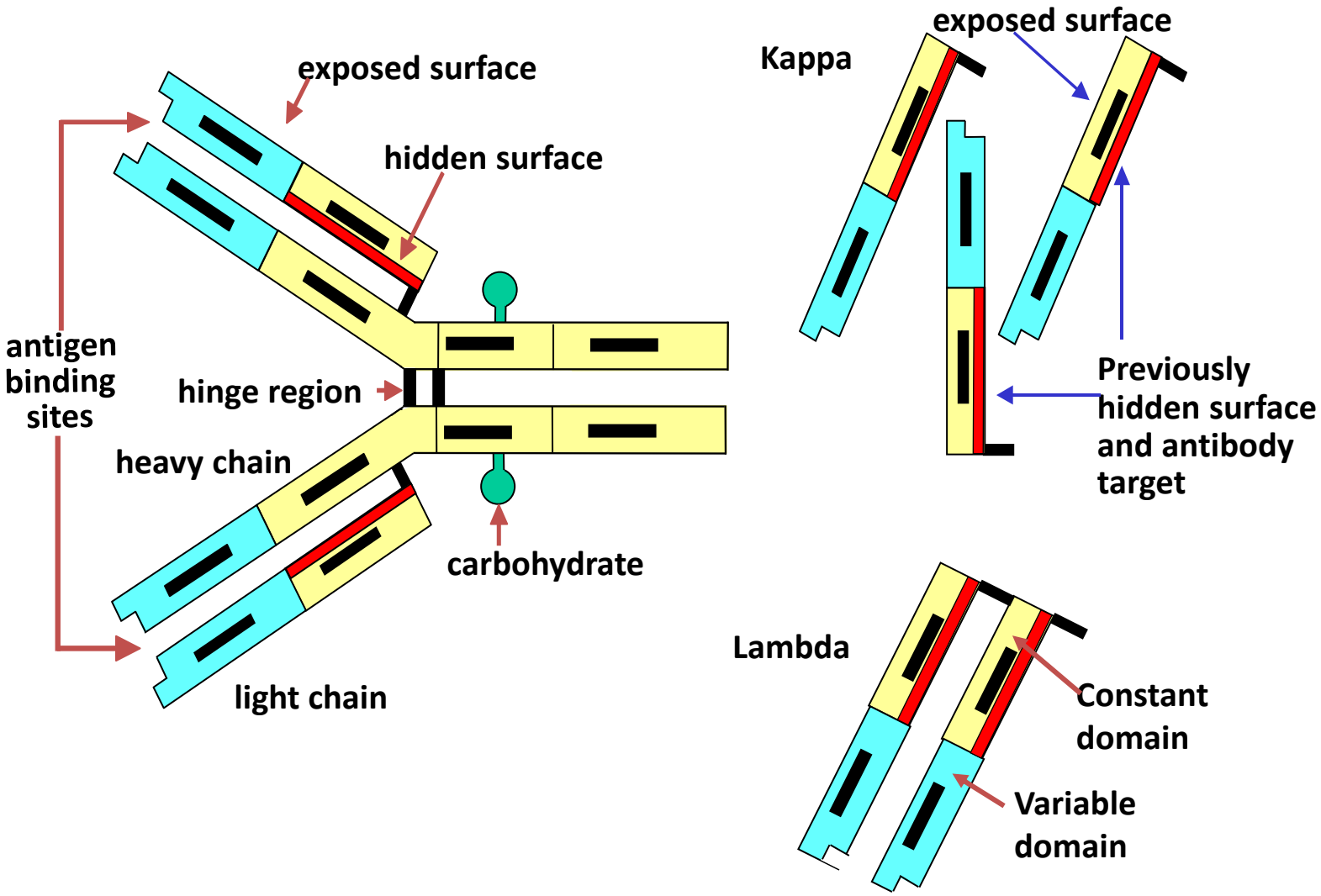
72 year old female presents with tiredness and hip pain

- Hb 110 (115-170), MCV 97 (81-98) WCC 5 (4-11), Plts 205 (150-450)
- Creat 123 (50-89), Corrected Calcium 2.95 (2.1-2.6)
- Low immunoglobulin's but no paraprotein detected

- Serum Free Light chains
  - Kappa 1004.0 (3.3 - 19.4)
  - Lambda 5.9 (5.7 - 26.3)
  - Ratio 170.2 (0.26 - 1.65)







An antibody molecule showing the heavy and light chain structure, together with free κ and λ FLCs.

Myeloma

- Can have paraprotein, light chain myeloma (20%), non-secretory myeloma (2%).
- 90% of myeloma has immunoparesis

- Prognosis 5-7 years
- MGUS
  - Common (10% over 80 yrs)
  - 1% per year chance of progression

CASE 6

64 year old female presents with lethargy, blurred vision and mild nose-bleed

Hb 109 MCV 90 WCC 4.0 Plts 156 (coag prolonged APTT)

U and E: normal

LFTs: bilirubin 7 (<21), Alk phos 63 (10 – 50), ALT 21 (10 – 50), Total protein 115 (60 – 80), Albumin 35 (35 – 50), Globulin 76 (22 – 36)

IgM kappa PP- 45 g/l

Waldenstroms/lymphoplasmacytoid lymphoma

- Low grade Non-Hodgkins lymphoma usually associated with a IgG PP

- Urgent marrow and CT
- Urgent plasma exchange
- Cyclophosphamide/steroid
- Rituximab later

CASE 7

72 yr old male bloods pre-op for Dupuytren's contracture.

Hb 134 (130-170) MCV 95 (81-98) WCC 11 (4-11) Plts 605 (150-450)

Repeat FBC was similar

Haematinics normal, CRP <10

JAK2 mutation=negative

CASE 8

57 year old female presenting with headaches

Hb 171 (115-170) MCV 89 (81-98) Hct 0.59 (0.39-0.45) WCC 12 (4-11) Plts 432 (150-150)

No meds

Lifelong smoker

BMI =20

JAK2 mutation=positive

U/S abdo: normal renal tract/ spleen 17cm

Myeloproliferative Disorders

- Essential thrombocytosis
  - Exclude iron deficiency/reactive causes
  - JAK2 (50%), CALR (20%), MPL (<10%)
  - Risk stratify and tx accordingly (aiming plts less 450 in pts >60)
- Polycythamia Rubra Vera
  - Relative/secondary (diuretics, sleep apnoea, RCC, lung disease)
  - Primary (JAK2 +ve 95%) (aiming Hct less than 0.45)
- Myelofibrosis

CASE 8

22 year old British Pakistani female

Well patient, picked up when going to donate blood

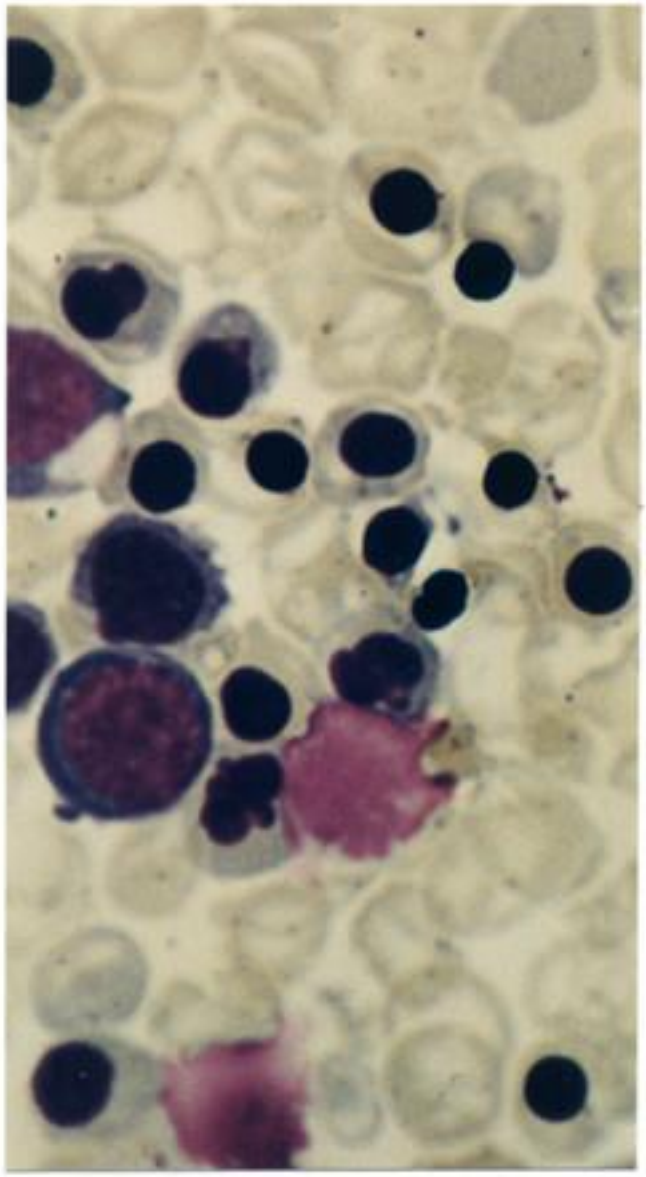
Hb 120 (115-170) MCV 65 (81-98) MCH 21 (27-33) WCC 5 (4-11) Plts 190 (150-450)

Ferritin 210 (30-400)

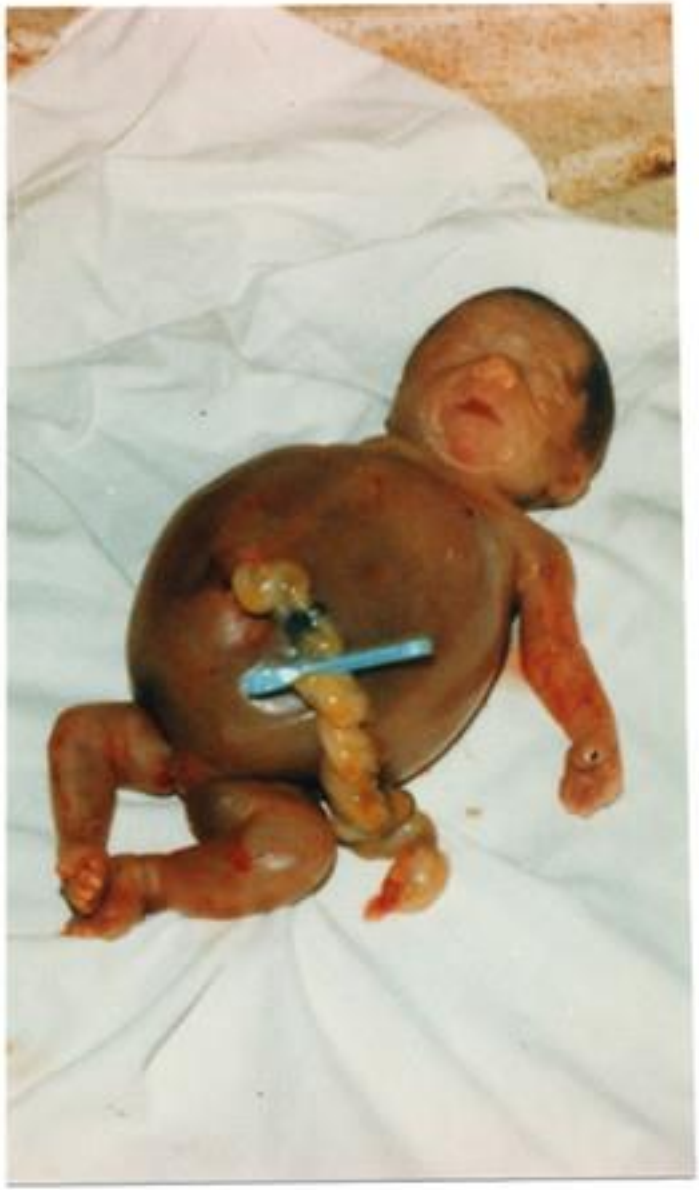
α thalassaemia

- 4 α genes per cell, normal (αα/αα)
- Silent α thalassaemia (-α/αα)
  - Very mild decrease in MCV and MCH in some
- α thalassaemia trait (--/αα or-α/-α)
  - Low MCV and MCH, mild anaemia in some
- Haemoglobin H disease (--/-α)
  - Hb 8-9, MCV and MCH low, hepatosplenomeagly, jaundice
- Haemoglobin Bart's hydrops fetalis (--/--)

a.



b.



- Not a clinical problem to patient
- Risks to future children
  - If father is also α thal carriers
  - Or carrier of other haemoglobinopathies
- Consider pre-conceptual testing
  - Screening partner (FBC/HPLC)
  - May need DNA testing
- Hb electrophoresis only helpful if normal iron stores, doesn't pick up alpha thal, diagnosis of exclusion then DNA tests.
- Nicole Paterson (sickle and thal nurse specialist) 0117 3422774

UHBristol Haematology

- URGENT- Haem SPR bleep 2445 or 2677 (in hours) (via switch out of hours)
- 2WW
- e-referral system (non urgent referral/advice)
  - Advice email (answer in 3 working days)
  - Referal to be seen
- Write Liaison Haematology service, BHOC, Horfield rd, BS2 8ED (or named consultant)
- Avoid fax, letters to haematology lab

Useful links

http://www.clinician.bristol.nhs.uk/clinician_portal/referral_support_tool.aspx

http://www.bcshguidelines.com/4_HAEMATOLOGY_GUIDELINES.html