King's College Hospital NHS
NHS Foundation Trust

The Paediatric Haemoglobinopathy Service at King's College Hospital

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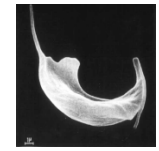

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The Haemoglobinopathies

Refers to a range of genetically inherited disorders of the red blood cell

Commonest forms include

- Sickle Cell Disease
- Alpha & Beta Thalassaemia

Occur commonly in populations whose ancestors come from Africa, Asia, Mediterranean Islands, middle & Far East

Autosomal recessive mode of Inheritance – Both Parents Have to be carriers

Frequency of Haemoglobin Trait (Carriers) in Ethnic Groups

Beta Thalassaemia Trait

- 1 in 7 Greek
- 1 in 10-20 Asian
- 1 in 12 Turkish
- 1 in 50 African and African Caribbean
- 1 in 1 000 English

Sickle Cell Trait

- 1 in 4 West African
- 1 in 50 African Caribbean
- 1 in 50 Asian
- 1 in 100 Northern Greek

- Worldwide Alpha Thalassaemia carrier states are More common than Beta Thalassaemia
- It is thought that 3% of the worlds population carry a thalassaemis gene

Thalassaemia

The most common Haemoglobin type is HbA and contains four globin chains

2 Alpha (α)

2 Beta (β)

Haemoglobin is inherited from both parents

4 Alpha genes (2 form each parent) αα/αα

2 beta genes (1 from each parent) β^ β^

The Thalassaemias are a group of disorders caused by the under production of one or more of the chains (α or β) that make up haemoglobin

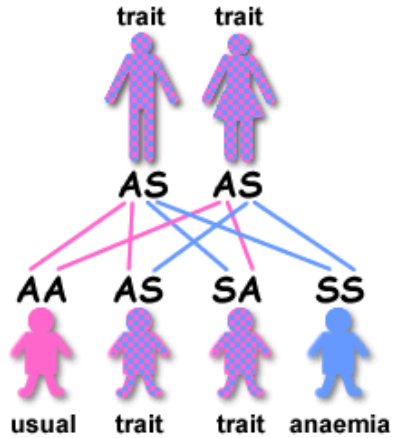
Unlike Sickle Cell Disease and other abnormal haemoglobins Thalassaemia effects the quantity of the globin chain that is synthesised

Sickle Cell Disease

Mutation of the beta gene affect the quality of the beta chain produced

Beta globin chain – 6th Amino Acid in Haemoglobin A - Glutamic Acid is substituted for Valine

Inheritance of Sickle Cell Disease



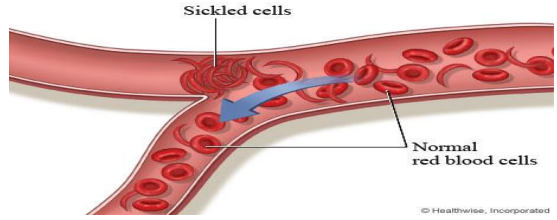
Sickle Cell Disease

Worldwide there are over 300 variants haemoglobin

Types common in the UK

- Haemoglobin SS (HbSS)
- Haemoglobin SC (HbSC)
- Haemoglobin S/β Thalassaemia (HbS/β Thal) (β+ or βº)

In Sickle Cell Disease, there is a sickle shaped abnormality of the red blood cells. These cells acquire a crystal like formation when exposed to low oxygen tension



Demography

In The UK there are approximately:

12 000 individuals with Sickle Cell Disease

600 with Beta Thalassaemia

Numbers increasing

- Advances in medical management
- Aging population – Increased life expectancy

  1973 – Average life expectancy 14 years

  HbSS – Men 42, Women 48 years. HbSC – Men 60, Women 68

  Some parts of the world halh these children die before their first birthday
- Population movement

Local Demography

Approximately 6 000 individuals with Sickle Cell Disease live within the South East Thames area – Largest group in The UK and Europe

King’s College Hospital
2 000 patients listed

On active register
400-500 Children

Thalassaemia – 25 paediatric patients with α & β Thalassaemia

Haemoglobinopathy Team – Paediatrics

Consultant – Haematology
Consultant – Haematology
Consultant – Haematology
Clinical Fellow
Specialist Registrar – Haematology (six month rotation)
2 Paediatric Clinical Nurse Specialists – Haemoglobinopathies
Clinical Psychologist – Haemoglobinopathies
Community Specialist Nurse – Haemoglobinopathies
Transition Specialist Nurse - Haemoglobinopathies

Acute Complications

- Vaso-Occlusion – Acute painful episodes
- Acute Chest Syndrome
- Stroke
- Infection
- Haemolysis
- Sequestration
- Aplastic

Multi-system disorder, causing chronic organ damage

Compounding factor – Disease is relatively unpredictable

Local Data

King's College Hospital – Largest centre for Sickle Cell Disease in The UK

Children's Services

- Over 1 000 children seen in outpatient clinics per year
- 150 inpatients per year
- 300 daycases
- Links with PICU – Referrals form other hospitals

Clinical Psychology

- Neuropsychometric Testing
- Cognitive behavioural Therapy

Outpatients

- Longest standing Sickle Cell & Thalassaemia clinic in the country
- Designated staff in paediatric Sickle Cell & Thalassaemia – Referrals from all over London & SE England
- Stroke prevention – Using TCD & MRI – Unavailable elsewhere
- Adolescent/Transition Clinic
- Joint Neurology Clinic

Day Cases

Phillip Isaacs - 9 Bedded ward

- Blood Exchange/Transfusion
- Regular Transfusion Programme
- Reviews
- Preparation for surgery at other hospitals

Children's Emergency Department

Date: Time: Clerked by:
Designation:

Name M/F

Dob: Age: Hospital number:

Haemoglobin Phenotype HbSS HbSC Hb S/β$^+$ Thal Hb S/β^0 Thal other:
G6PD
Steady state Hb g/dl Bilirubin µmol/L

Medication: Penicillin prophylaxis Y/N
Usual analgesia Ibuprofen mg
Paracetamol mg
Codeine Phosphate mg
Other mg

Past medical history:
Stroke Y/N Acute chest syndrome Y/N admission for pain Y/N
Parvovirus Y/N Acute splenic sequestration Y/N biliary colic Y/N
Surgical history

Relevant social issues

Presenting complaint

Red flag (see cliniweb)
Severe pallor
Severe jaundice
Temp> 38.5°C
Tachypnoea
Chest pain
Severe pain esp abdomen
Weakness without pain
Parasthesiae
Altered consciousness
Priapism

If pain
Site:
Analgesics taken at home Paracetamol mg at h
Ibuprofen mg at h
Codeine phosphate mg at h

Haemocue: CXR
Hb
WBC
PLT
Bilirubin Oxygen sats in air
Urea

General findings ENT/ Lymphnodes
Alertness
Distress
Pallor
Jaundice

CVS/RS
Cardiomegaly
Heart sounds
Murmurs
Tachpnoea
Nasal flaring
Air entry
Added sounds

AS
Tenderness
Spleen size
Liver size
Distension
Bowel sounds

CNS
Cranials
Fundi
Power
Tone
Reflexes
Asymmetry

Pain /swelling/tenderness

If unwell or in pain
- Ensure IV access
- Keep warm
- Maintain hydration
- Give pain relief

Pain score at h after

Diagnosis/impression Admit : *unwell <1yr*
Temp>38.5
Hb< 5g or <2g below steady state
Neurological signs/symptoms

Plan:

Investigations

Treatment:

Admit/ Discharge

A copy of this document must be faxed to S E London Sickle cell & Thalassaemia centre 0203 049 6085

Inpatient Wards

Toni & Guy Ward – Medical

Children's Surgical Ward – General & Neurosurgical

Rays of Sunshine Ward

Thomas Cook Children's Critical Care Centre PICU & CHDU

- Acute admissions
- Coordination of surgical admissions
- Admissions under other specialist teams eg. Liver, Neuro
- Admissions to critical care unit

Paediatric Short Stay Unit

Paediatric Short Stay Unit - PSSU

6 Bedded unit situated in the Emergency Department, opened in 2014

- Acute admissions >48 hours
- Rapid Response Clinic
- Hospital @ Home Service

Role of The Haemoglobinopathy Team

- Supporting families
- Ongoing Health Promotion
- Workshops
- Support Groups
- Staff Training/Education
- Liasing with other organisations
- Patient literature
- Protocols/guidelines
- Clinical Audit & Research

Thank You

Any Questions?