correction factor of 3; for platelets, counts of 20–30 or as specified ie >50 post neurosurgery, or >75 for LP; and for thalassemia Hb. >9g%. We also used as an estimate volume in a Red cell unit as 270mls and platelet unit as 180mls, unless specified.

Results

Appropriate in time:

Thalassemia: 20/20 Red cell transfusions were all elective and in routine hours.

Haem-Onc.: 40/60 were elective of which 34/40 occurred in routine hours. 18/60 were "pragmatic" ie. in anticipation of chemo, procedures, sepsis, discharge etc., of which 7/18 occurred in routine hours. 2/60 were urgent transfusions, occurred in routine hours.

Appropriate in amount:

The weights across all 10 patients ranged from 8 – 49kg.

Abstract G182(P) Table 1

Patient Group		Mean	Median	Range
Haem-Onc. Red cell transfusions	Pre transfusion Hb (G%)	7.6	8	4.6–8.5
	Volume (ml/kg)	11.9	12	5.5–20
	Post transfusion Hb (G%)	10.9	11.2	8.2–13
Haem-Onc. Platelets	Pre transfusion count	38.5	40.5	14–56
	Volume (ml/kg)	7	4.1	3.8–16.7
	Post transfusion count	85	68	52–388
Thalassemia Red cell transfusions	Pre transfusion (G%)	9.1	9.1	8.3–11
	Volume transfused (ml/kg)	17.1	17.6	14–23

Conclusions 85% elective and 40% pragmatic transfusions occurred in routine hours. Pre transfusion thresholds and volume of transfusions in all 3 groups were acceptable. The variation in prescribed volumes reflects minimising exposure and wastage by prescribing in units wherever possible. These correlations are best visualised on the attached slides.

G183(P) COST EFFECTIVENESS OF HYDROXYUREA THERAPY IN PREVENTING INPATIENT ADMISSIONS DUE TO VASO-OCCLUSIVE CRISIS IN SICKLE CELL CHILDREN – A DISTRICT GENERAL HOSPITAL EXPERIENCE

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Aim To study the effectiveness of hydroxyurea in reducing the number of inpatient admissions due to vaso-occlusive crisis in children with sickle cell disease and estimation of potential cost savings for the NHS.

Methods Retrospective audit of number of admissions, length of stay, use of intravenous opioids over a period of 2 years before and after starting hydroxyurea treatment in 5 children with sickle cell disease.

Results Of 5 patients with sickle cell disease 3 with HbSS and 2 with HbSC were started on hydroxyurea over the last 5 year period. Comparison was made between the numbers, severity and duration of the vaso-occlusive episodes needing inpatient admission before and during treatment are made and potential cost savings for the NHS estimated.

After starting treatment with hydroxyurea there was a reduction in the number of inpatient admission by 38% (50 vs 31) and inpatient days by 43%(184 Vs 105). A reduction in proportion of admissions needing iv opioids (20% Vs 12.9%) as well as in the number of inpatient days on iv opioids (29 days vs 9 days, reduction by 69%). In one patient the treatment was stopped after 11 months due to poor compliance with blood tests. All patients showed decrease in the number of admissions and opioid use except for one patient in whom there was an apparent increase in both. The diagnosis was revised as sickle cell with fibromyalgia syndrome and hydroxyurea was later discontinued without increase in pain symptoms.

Total cost of hydroxyurea treatment over 2 year period for the 5 patients was £3602. There was a reduction in number of high dependency admission by 20 days and non-high dependency admission day by 60. The total potential cost saving from reduced admission was £73220. Net savings for the NHS estimated over the 2 years since starting hydroxyurea for the above 5 patients was £69618.

Conclusion Commencing hydroxyurea treatment after careful patient selection in sickle cell children with frequent vaso-occlusive crisis can achieve significant cost savings for the NHS while improving the quality of patient care.

G184(P) THROMBOPHILIA TESTING IN CHILDREN; THREE CASE VIGNETTES

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Aim An increasing number of children are being tested for inherited thrombophilias. We present three cases of children known to our centre, who were tested for and found to have a positive thrombophilia test to illustrate the wide spectrum of issues and challenges that can arise and the implications for the children, their families and the health care professionals involved in their care.

Methods In most children a combination of risk factors lead to the development of symptomatic VTEs, and idiopathic VTEs, unlike in the adult population are extremely rare.

Thrombophilia testing in children with a proven VTE, is controversial, and at present there are no guidelines for thromboprophylaxis in children with heritable thrombophilia. Even more challenging is testing children who are asymptomatic. Clearly the family history of the inherited thrombophilia is relevant, but also of importance is the knowledge that any genetic testing should be done with the purpose of improving outcome for the individual concerned.

Results The three chosen examples are children aged between 2 and 16. The first case is a young girl with an extensive family history of childhood thrombosis in her father and uncle, who has inherited the same protein C mutation but is asymptomatic herself, the second is a young boy with a family history of protein S deficiency who was diagnosed with a PE and found to have the same mutation. And the third is a young boy who was tested inadvertently but was found to have antithrombin deficiency.

Conclusions These are only three examples from our haemostasis & thrombosis centre chosen to illustrate some of the challenges that arise in testing children for inherited thrombophilias and their long-term management and follow up. There is very little information available with regards to thrombophilia testing in asymptomatic children, which highlights even more so the importance of ensuring that the child and their family are reviewed by specialists in coagulation medicine and the decision to test for inherited thrombophilia is made based on the information obtained in the history, relevant to that child with appropriate counselling prior to testing and with a knowledge of the limitations of thrombophilia testing.

G185(P) FEVER AND BACTERIAL INFECTIONS IN CHILDREN WITH SICKLE CELL DISEASE

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Background and aims Children with sickle cell disease (SCD) are at an increased risk of developing bacteraemia and other serious bacterial infections, which can be associated with significant morbidity and mortality. Fever, however, is a common symptom in children with

SCD and can occur with both infection (bacterial and viral,) and sickle cell crises. This study aimed to look at the incidence of bacteraemia and bacterial infections in children with SCD presenting to a North-East London district hospital with a fever of 38.5 degrees or higher.

Methods A retrospective analysis was performed on all children (aged under 16 years,) with SCD presenting with a fever of 38.5°C or higher over a 1-year period. Data was collected for each febrile episode on age of child, type of SCD, final clinical diagnosis, initial White cell count (WCC) and C-reactive protein (CRP) levels, blood culture and microbiology results, length of stay and clinical outcome. Children were divided into those having a definite bacterial infection, suspected bacterial infection (clinically suspected but no microbiological confirmation,) or no bacterial infection. Definite bacterial infection was defined as bacteraemia (the isolation of a non-contaminant bacterial from the blood culture,) or other bacterial infection with positive microbiological confirmation.

Results Over the 1-year period there were 88 episodes analysed in 59 children. Definite bacterial infection occurred in 8% of febrile episodes of which 3.4% had bacteraemia. (Streptococcus pneumonia, Salmonella hartford, Salmonella typhirium.) Suspected bacterial infection occurred in a further 33% of episodes. In 59% of episodes the final diagnosis was either a sickle cell crisis or viral illness (no bacterial infection.) Diagnosis did not vary significantly by haemaglobinopathy. One death occurred from Salmonella typhirium septicaemia. Average length of stay varied from 3.6 days in the group with no bacterial infection to 8.9 days in the group with definite bacterial infection.

Conclusion Bacterial infections continue to be a significant problem in children with sickle cell disease. Salmonella infection is a growing concern in this group of children. Further work is required to identify risk factors and predictors for bacterial infection, and ascertain optimal prevention and management strategies.

G186(P) AN AUDIT OF MANAGEMENT OF PAIN CRISES IN CHILDREN WITH SICKLE CELL DISEASE

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Aims Sickle cell disease (SCD) is a lifelong haematological disorder resulting in anaemia and pain crisis. Specialist centres use experienced staff and accredited protocols to manage pain crises in affected children. The main objective of this audit is to identify areas of management of crises which could be improved in line with recommendations set out locally, by the 'Painful Sickle Cell Crisis Protocol' and nationally, by the NHS Sickle Cell disease in Childhood Standards and Guidelines for Clinical Care. This may provide useful insight for service improvement and evidence for similar units throughout the UK and Europe. Of particular interest was timing and efficacy of administration of analgesia.

Methods Baseline audit of management of patients presenting to Accident & Emergency (A&E) or the hospital's haematology day unit (DU) due to acute pain crisis of SCD was conducted. The study cohort was all patients seen at the hospital with a diagnosis of SCD from 1st January 2010 to 31st December 2011. 43 patients met this criteria. Re-audit to be done January 2014.

Results Of the 43 patients registered with the hospital haematology unit, 12 attended the hospital because of pain crises, whilst 31 had no pain crises within the allotted time frame. 5 patients experienced more than one acute pain crisis. Time to administer initial analgesia was 57 ± 37 minutes. Average length of stay in hospital was 4.5 ± 3 days. The most common initial analgesic administered was oral or IV morphine. 14 of the 25 children had tried medication at home prior to presenting.

Conclusions Aspects of the service identified for improvement include: clear documentation of time patient presents, time they are seen by a doctor and time they are given their first analgesia; unequivocal inclusion of a drug kardex in patients notes and consistent use of a pain scoring system.

Recommendations Implementation of a 'Sickle Cell Pain Crisis Assessment and Management' form. This would improve consistency of documentation of information relating to pain crises and therefore accuracy of future service monitoring.

G187(P) CAUSES OF SEVERE ANAEMIA (HB <5 G/DL) IN CHILDREN (<18 YEARS) BETWEEN 2006 AND 2009

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Severe childhood anaemia risks significant morbidity and mortality though may have different benign or malignant aetiologies. We set out to define the characteristics of this group of patients at our institution.

This study was performed in a large paediatric teaching hospital. We retrospectively identified children (<18y) presenting with Hb <5g/dL in the period 2006–9 by use of computerised laboratory records. Case notes were then reviewed and we recorded patient characteristics, final diagnosis and management with 2y of follow-up.

93 patients fitted study criteria. Patients were mean age 5y, (range 1d-17y). Diagnoses were: 33/93 leukaemia (25/33 acute lymphoblastic, 7/33 acute myeloid, 1/33 juvenile myelomonocytic leukaemia); 23/93 iron deficiency anaemia (IDA); 12/93 hereditary blood disorders (including 7/12 hereditary spherocytosis with 6/12 associated parvovirus); 25/93 "other" including 6/25 haemolytic-uraemic syndrome and 3/25 transient erythroblastopaenia. Of leukaemia 5/33 had presenting white cell count > $100x10^9$/L, and all received red-cell transfusion. Of IDA 19/23 had nutritional IDA (nIDA). 17/19 nIDA were aged <3y. 11/17 nIDA were of Pakistani origin (versus 3.5% of city population). Linking residential postcode with national index of multiple deprivation, 11/17 nIDA lived in the most (lowest 20%) deprived areas, rising to 9/11 in more severe nIDA (Hb <4g/dL). In IDA, all were prescribed iron supplementation, 21/23 feeding practises reviewed by dietician and 17/23 were transfused red-cells (all those with Hb <4g/dL). 9/23 IDA resolved within 1y though 3/23 had no repeat Hb recorded. Of all patients 11/93 died: 3/11 at initial presentation, 6/11 within 1y and 2/11 within 2y.

We show that severe anaemia is most commonly caused by acute leukaemia in this population. However nIDA due to poor infant feeding practise is an important preventable diagnosis and may merit particular health education. A disproportionate number of such patients are from deprived areas. A significant number of patients with nIDA had persistent anaemia >1y post presentation suggesting the need for structured follow-up and ongoing intervention.

British Society of Paediatric Gastroenterology, Hepatology and Nutrition/British Paediatric Neurology Association

G188 THE ROLE OF SCREENING INVESTIGATIONS IN CHILDHOOD ARTERIAL ISCHAEMIC STROKE

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Childhood arterial ischaemic stroke (AIS) is a heterogeneous disorder, with morbidity in 2/3rd of survivors and recurrence in 10%. Current clinical guidelines recommend a wide range of investigations