CHARACTERISING THE DISEASE PRESENTATIONS OF LCH OVER 10 YR AT A UK PTC

1AFM Mitchell, 2R Ramanujachar, 3J Gray, 4G Nicolin, 5U Uparkhar, 7B Vadgama, 1A Cooke. 1Paediatric Oncology, Southampton Childrens Hospital, Southampton, UK; 2Paediatric Oncology, University of Southampton, Southampton, UK; 3Paediatric Pathology, University Hospital Southampton NHS Foundation Trust, Southampton, UK

Background Langerhans Cell Histiocytosis (LCH) describes a clinical spectrum of disease caused by accumulation of pathologicalangerhan’s cells infiltrating tissues. Further understanding regarding the patho-physiology and natural history of langerhans cell histiocytosis has emerged with the understanding of its status as a clonal BRAF stimulated malignancy.

Objectives To characterise the disease presentations of Langerhans Cell Histiocytosis in a UK Principal treatment centre (PTC) over the past 10 years.

Methods Our PTC serves a population of 3.6 million sharing with 11 regional shared care centres. A prospective case review was performed for all patients treated for LCH over the last 10 years. Data were collected on demographics, disease location, treatment modality, intervals of remission free outcome and latest evaluation at follow up.

Results 1041 children have been treated for cancerover the past 10 years (1/1/2006–31/12/2016). We identified 45 cases of LCH from children aged 2 mths to 16 years (median age 9 years (range, 5–15), M:F 3:1 were randomised into RUTF (37) :control(33) arms. Median protein and calorie intake as well as weight gain at 6 weeks(2.6 kg vs. 2 kg) was higher in the RUTF arm compared to controls on standard dietary care. At 6 weeks there was significant increment of lean mass in the RUTF arm vs. controls. Vitamin B12 and folate deficiency (33%), vitamin D(63%), 56% and 96% had copper and zinc deficiency respectively, which improved in the RUTF arm. Children on RUTF experienced significant reduction in the episodes of febrile neutropenia (18.9% vs. 30.3%, p<0.06), protocol delays(2.7% vs 30.3%, p<0.05), grade 3/4 neutropenia (40.5% vs. 66.7%, p<0.05), thrombocytopenia(21.6% vs. 30.3%, p<0.05) and anaemia (18.9% vs. 36.36%, p>0.05) beyond 6 weeks. Mean HRQOL scores were better in the RUTF arm at baseline, 6 weeks and 3 months.

Conclusion RUTF is cost-effective in improving nutritional status resulting in higher weight and lean body mass which translates into improved treatment tolerance and QOL.
Aims We aimed to conduct a case series review of severe sepsis episodes in patients with sickle cell disease at a large regional paediatric centre in the UK. We aimed to examine sickle cell disease prior to episode, compliance with sepsis prevention, type of infection including the organism isolated, sequelae, and outcome.

Methods Retrospective cumulative case series of severe sepsis admissions over the last 5 years among our cohort of patients with sickle cell disease at a large paediatric regional centre in the UK. Data was extracted from the department’s sickle cell database and from clinical records.

Results We present 5 cases of severe sepsis among our regional cohort of 388 children with sickle cell disease. The patients presented within a 2-year period between May 2015 and May 2017. The ages of the patients ranged from 2 to 11 (median age 3 years). It was the first admission to hospital in 3/5 cases. One patient was on hydroxyurea and had received childhood vaccines, and 3/5 had received the polysaccharide pneumococcal vaccine. The organisms isolated were *Strep. pneumoniae* (3 cases), *E. coli* (1 case), and *Salmonella durham* (1 case). Sepsis was often rapidly progressive and presented atypically. Two of the 5 patients died following sepsis episode. Both had pneumococcal sepsis. There was significant morbidity in survivors. Complications included osteomyelitis (2 cases), pathological fracture (1 case), hearing impairment and central diabetes insipidus secondarily to meningoencephalitis (1 case) and necrosis of the fingertips. In the pneumococcal cases, either patient was not covered with vaccination, or the organism had partial resistance to penicillin.

Conclusion Despite advances in recent years following introduction of sepsis prevention measures, sepsis remains an important cause of mortality and morbidity in paediatric patients with sickle cell disease. Sepsis often presents rapidly and atypically in this group of patients.

Background and aims Allogeneic haematopoietic stem cell transplantation (HSCT) is widely used to treat non-malignant conditions. Mixed chimerism (MC) is an increasingly observed phenomenon in such cases. This study’s purpose was to explore predictors of MC and graft failure, variations in outcome between patients with complete chimerism (CC) and MC, and the utility of lineage-specific chimerism in predicting graft outcomes.

Methods Our patient sample included 284 HSCTs performed in children with non-malignant conditions between July 2000 and March 2017 at our centre. The following variables were considered in each patient: gender, age at transplant, date of transplant, disease, conditioning regimen, T-cell depletion, donor and stem cell source, alive/deceased status, chimerism status. Variables were assessed using univariate and multivariate logistic regression analysis. The relationship between myeloid...