TFT were normal. Bone marrow biopsy revealed mild aplasia, and no myelodysplastic syndrome or leukaemia. HIV, Hepatitis screen and viral serology were negative. Fanconi Anaemia test came back positive.

Conclusions Children presenting with macrocytosis should be carefully evaluated and referred to a haematologist, as it could be the first manifestation of a serious underlying bone marrow problem.

Macrocytosis in age range of six months to 12 years age is MCV >90.

Causes of macrocytosis include disorders of folate/B12 metabolism, liver disease, congenital heart disease, Down's syndrome, hypothyroidism, drugs (anticonvulsants, zidovudine, hydroxyurea, immunosuppresants), myelodysplastic syndromes, bone marrow failure, and rare genetic syndromes (eg, Fanconi anaemia, thiamine-responsive megaloblastic anaemia syndrome).

G408(P) CYTOKINE STORM ASSOCIATED MULTI-ORGAN FAILURE WITH POOR NEUROLOGICAL OUTCOME, DURING RITUXIMAB ADMINISTRATION IN A CHILD WITH RELAPSED ACUTE LYMPHOBLASTIC LEUKAEMIA AND EBV RELATED LYMPHOPROLIFERATIVE DISEASE

[1]C Kanaris, [2]RF Wynn, [3]A Konstantinidis. *[1]Paediatric Intensive Care Unit, Royal Manchester Children's Hospital, Manchester, UK; [2]Bone Marrow Transplant Unit, Royal Manchester Children's Hospital, Manchester, UK; [3]Paediatric Haematology Department, Royal Manchester Children's Hospital, Manchester, UK*

10.1136/archdischild-2015-308599.362

We present the case of a 6 year old boy, with relapsed Acute Lymphoblastic Leukaemia and subsequent matched unrelated bone marrow transplant. His Epstein Barr Virus (EBV) titres were significant 6 months post-transplant with evidence of EBV related lymphoproliferative disease. Following local protocols, Rituximab was used for therapy and within minutes of starting the infusion the patient suffered pyrexia, hypotension and seizures with subsequent multi-organ failure, due to a cytokine storm. Organ damage included fulminant hepatic failure, renal failure meriting dialysis, cardiac failure requiring inotropic support and a significant acute brain injury. There was significant neurological impairment with radiological and electrophysiological evidence of global brain cell damage. As a result of irreversible neurological injury, the focus of treatment was switched from curative to palliative.

Cytokine storm is an immune mediated phenomenon, characterised by an overwhelming release of cytokines. This can produce a sepsis like response and may lead to multi-organ failure. Cytokines are normally produced by leucocytes as a response to an infective or inflammatory process and their main role is to induce leukopoeisis.

Rituximab is an anti-CD20 monoclonal antibody that has been useful in treating EBV related lymphoproliferative disease. Rituximab triggers the rapid release of cytokines that may lead to a cytokine storm with maximum levels within 2 h from the start of the infusion.

Cytokine storms are characterised by overproduction of immune mediators that in turn lead to cellular overactivation, increased endothelial permeability, polymorphonuclear neutrophil adhesion and migration. As there is lack of regulatory intervention, tissue congestion with activated leucocytes ensues which eventually causes parenchymal injury. Although we report multi-organ failure in this case, lungs and gut are the organs that are more commonly affected.

The incidence of cytokine storm in paediatric oncology patients is not known. It has been associated with haemophagocytic lymphohistiocytosis, graft-versus-host disease following haemopoetic stem cell transplant and the use of certain monoclonal antibodies. As in this case, results of the cytokine storm can be catastrophic, despite early recognition.

G409(P) AN EVALUATION OF THE TRANSITION TO ADULT CARE FOR YOUNG PATIENTS WITH SICKLE CELL DISEASE

[1]H Sivaguru, [2]S Mohun Kemp, [2]R Crowley, [2]G Hann, [2]DA Yardumian, [2]M Roberts-Harewood, [2]O Wilkey. *[1]University College London, London, UK; [2]North Middlesex University Hospital, London, UK*

10.1136/archdischild-2015-308599.363

Aims More than 95% of children with sickle cell disease (SCD) survive into adulthood, making a successful transition from child to adult care an essential process. Poor management of this transition may result in reduced compliance, high non-attendance rates and adverse effects in later life. This study aimed to investigate the views of young patients in the process of transitioning with the long-term goal of improving previously poor engagement within the adolescent population.

Methods A questionnaire was given to all SCD patients between the ages of 13 and 21 who attended out-patient clinics over a four-month period.

Results 31 questionnaire responses were collected (response rate 94%).

Overall satisfaction with the transition process varied with age. The mean score was 7.4/10 among 16–21 year olds, but only 4/10 among 13–15 year olds.

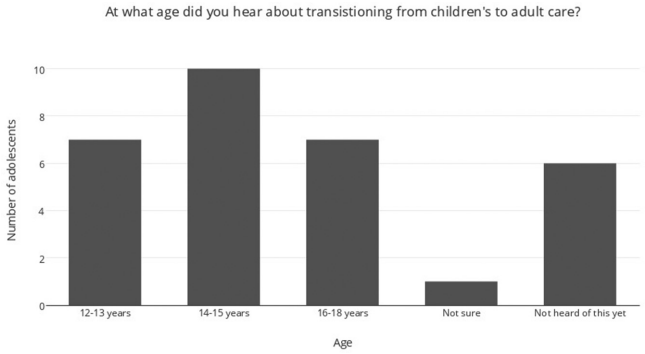
Praise was given for transition services such as a previously organised tour of the adult department, a peer discussion day and an information pack.

48.4% were interested in speaking to peers about their experiences.

41.9% reported receiving a transition information pack of which 81% found it useful.

Adolescents recounted that the topic of transitioning had been first broached at a wide range of ages (see Figure 1).

Conclusion There are discrepancies between the planned transition programme and reported patient experience. The tour of adult department and peer discussion day were praised, however not every adolescent recounted experiencing these services. Feedback from clinicians suggests that even though adolescents like the idea of these transition services they still fail to attend. A focus group has been initiated to improve adolescent



Abstract G409(P) Figure 1

participation. The information pack was introduced in 2010 which may explain why under half reported receiving it. Notably the majority of those that received it found it useful, suggesting that it is an important component of transition. The age that the topic of transition is introduced could be optimised and standardised, as it is widely thought that beginning the transition process early is an essential element for successful transfer to adult care.

G410(P) ARE ADOLESCENTS WITH SICKLE CELL DISEASE SATISFIED WITH THEIR OUT-PATIENT CLINIC EXPERIENCE?

[1]S Mohun Kemp, [2]H Sivaguru, [1]R Crowley, [1]DA Yardumian, [1]M Roberts-Harewood, [1]O Wilkey, [1]G Hann. *[1]North Middlesex University Hospital, London, UK; [2]University College London, London, UK*

10.1136/archdischild-2015-308599.364

Aims Adolescents with chronic conditions can prove difficult to engage. Each clinical encounter must be maximised to provide a positive experience that encourages continued attendance and a collaborative relationship between doctor and patient. This project investigated the out-patient experience of adolescents with sickle cell disease.

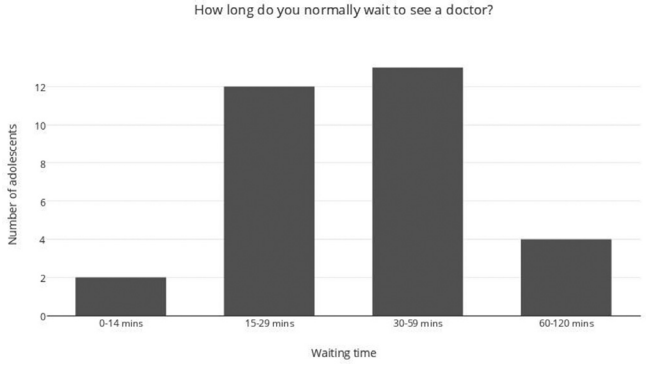
Methods A questionnaire comprising both qualitative and quantitative questions was distributed to patients aged 13 to 21 years who attended haematology clinics between January and April 2014.

Results 31 adolescent patients completed the questionnaire (response rate of 94%).

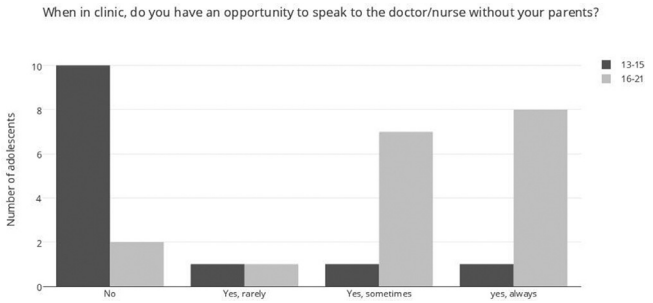
When asked to rate their out-patient experience, the mean score was 6.7/10. Qualitative questioning highlighted themes of dissatisfaction around long waiting times and lack of waiting room activities. There was a wide variation in reported waiting times (see Figure 1). There was a significant relationship between higher waiting times and lower overall out-patient experience score.

Positive comments were made about 'friendly and helpful' health professionals with a rating score of 8.1/10 for staff friendliness.

During the consultation the majority of patients felt they had enough time to discuss their concerns. Older patients were more likely to have the opportunity to speak to the doctor or nurse without a parent present (see Figure 2). Of those who did not have this opportunity, 23% stated they would like to talk to a doctor alone.



Abstract G410(P) Figure 1 Waiting times



Abstract G410(P) Figure 2 Discussion with doctor

Conclusion There is room to improve the out-patient clinic experience for patients with sickle cell disease. Specifically, reducing waiting times should be given priority. Both clinicians and patients need to arrive promptly to prevent a backlog delay. The clinic environment could be modified to provide a more comfortable and stimulating place for adolescents. Although financial and spatial constraints limit refurbishment, patient feedback suggests simple measures such as installing a water dispenser in the paediatric waiting room may improve patient satisfaction. Finally adolescent patients could be given more opportunity to speak to clinicians without a parent present and this could be introduced at a younger age. However, both our doctors and the adolescents themselves reported that there is often parental resistance to this, which needs to be overcome.

G411(P) A RARE CASE OF PRIMARY ANTIPHOSPHOLIPID SYNDROME

R Onita, C Reynolds, A Joshi. *Luton and Dunstable University Hospital, Luton, UK*

10.1136/archdischild-2015-308599.365

Introduction Antiphopholipid syndrome (APS) is a systemic autoimmune condition characterised by venous or arterial thrombosis, hypercoagulability and pregnancy comorbidities in the presence of circulating antibodies directed against phospholipids. Cerebrovascular disease including sinus vein thrombosis and ischaemic stroke are the presenting features in approximately 30% of cases. Primary APS in the paediatic population is very rare with the exact incidence unknown. A significant number of cases of APS will be associated with autoimmune disease, particularly systemic lupus erythematosus. There are no studies on the management of paediatric APS, which makes the management of these children a challenge for the paediatrician

Case description A 2 year old girl, with no comorbidities presented to the local hospital with one episode of tonic-clonic seizure. Of note, is that for the previous days she became more lethargic, decreased appetite and decreased speech. She was developing normally and there was no family history of note. She had a full septic screen but this showed no evidence of acute infection. An MRI head showed left transverse sinus thrombosis with bilateral thalamic infarcts and right basal ganglia infarction. In view of these findings she had a full thrombophilia screen. The results showed evidence of APS with positive anti cardiolipin antibodies. There was no laboratory features of a secondary autoimmune disease, so a diagnosis of primary APS was made (ANA, ANCA, R. F., C3, C4, IG A, G, M-, Direct Antiglobulin test, PR3, MPO,-negative). She has been anticoagulated with warfarin but her ongoing management has been difficult.

Discussion Evidence for the treatment of APS is based on adult studies and the treatment itself comes with it's own