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, immunosuppressants), myelodysplastic syndromes, bone marrow failure, and rare genetic syndromes (e.g., Fanconi anaemia, thiamine-responsive megaloblastic anaemia syndrome).

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 leukopoiesis.

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.

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.
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?**

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**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.

**G411(P)**

**A RARE CASE OF PRIMARY ANTIPHOSPHOLIPID SYNDROME**

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**Introduction** Antiphospholipid 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 paediatric 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...