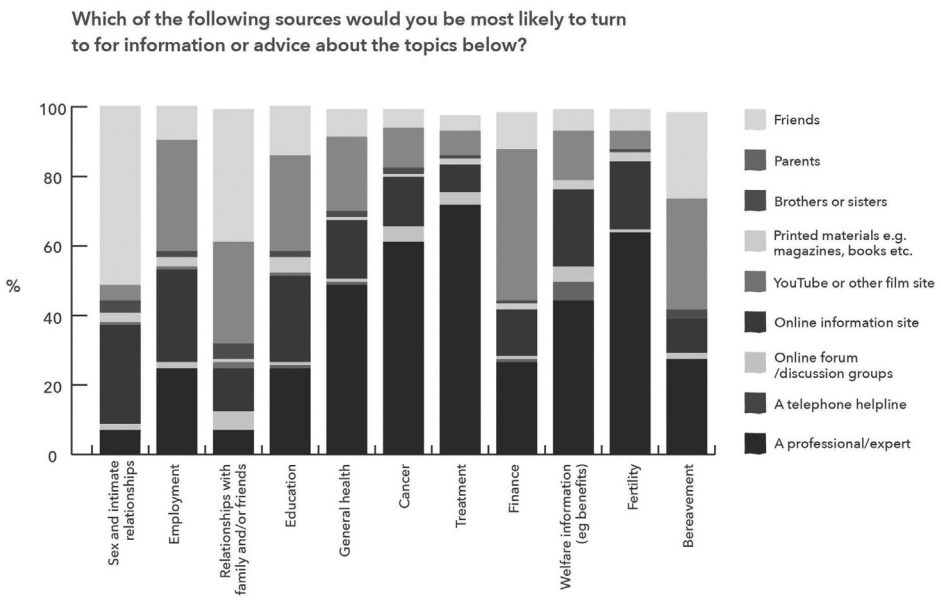

Abstract G406 Figure 1

resilience. To raise awareness among health and social care practitioners of young people's information and support needs.

Method

- A review of relevant literature was undertaken to understand the evidence base on help seeking and resilience and its applicability to young people with cancer.
- 138 young people with cancer were consulted through an online survey (124) and a focus group (14). All participants were self-selecting and identified through CLIC Sargent's networks. All had been diagnosed with cancer in the UK between the ages of 16 and 24, or were currently within that age range.
- Interviews were conducted with academics and practitioners to understand their views of help-seeking behaviour and resilience among young people with cancer.

Results

- Access to quality information is key to helping young people with cancer feel confident managing their illness and building resilience.
- Young people can feel anxious when their treatment ends and need more information and support at this point
- The source of information young people choose depends on the topic; 96% of survey respondents use the internet to look for information and advice for themselves; 74% of the young people would want to speak to a healthcare professional if they had a problem that was worrying them.
- Young people need a suite of support tools available to them.
- It's useful to conceptualise resilience as an outcome: the outcome of successfully coping with stressful experiences. 'Resilience' is often linked to 'coping' and 'competence'. Approaches to building resilience include those focused on individuals and those that take the wider health and social care system into account. Both approaches are relevant to young people with cancer.

Conclusion

- The research has enabled CLIC Sargent to reflect on its services and identify ways to improve support for young people. We also propose priorities for change for the wider health and social care sector, to move us all towards a system of support that fosters resilience in young people with health conditions such as cancer.

G407(P) MACROCYTOSIS – A RARE BUT SERIOUS PROBLEM!

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10.1136/archdischild-2015-308599.361

Aims A case report to highlight the importance of macrocytosis.
Methods A 7½ year old boy was referred with macrocytosis (MCV 108). He was fit and thriving child (weight, height 50th centile). History and examination was unremarkable. Bloods done 5 years ago also showed high MCV (95). Routine tests for evaluation of macrocytosis were normal. Later FBC showed thrombocytopenia, leucopenia and neutropenia. He was finally diagnosed to have Fanconi anaemia. He has been put on register for bone marrow transplant, and whole family is being tested for gene mutations.

Fanconi anaemia is an autosomal recessive or X-linked form of inherited aplastic anaemia. Sixteen mutations (FA-A to FA-Q) have been identified. Hematologic findings evolve over months to years, with 70% of children developing bone marrow failure by age 10. They are at high risk for developing myelodysplastic syndrome, acute myeloid leukaemia, and squamous cell carcinoma. Characteristic congenital malformations are present in up to 70% of affected children (short stature, café-au-lait spots, hypopigmented spots, thumb abnormality, microcephaly, hydrocephaly, hypogonadism, developmental delay). Diagnosis is made by presence of increased chromosomal breakage in lymphocytes cultured in the presence of DNA cross-linking agents (mitomycin C or diepoxybutane). Bone marrow transplant is the only available curative treatment. Other treatment options include androgens, hematopoietic growth factors, and blood product transfusions with supportive care.
Results Hb 122, WCC 8.6, Platelets 164, MCV 102 in 2011. Hb 118, WCC 5.4, Neutrophils 1.34, Platelets 130, MCV 108 in 2014. U&E, LFT, immunoglobulins, B12, Folate, Ferritin, and

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

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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

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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.

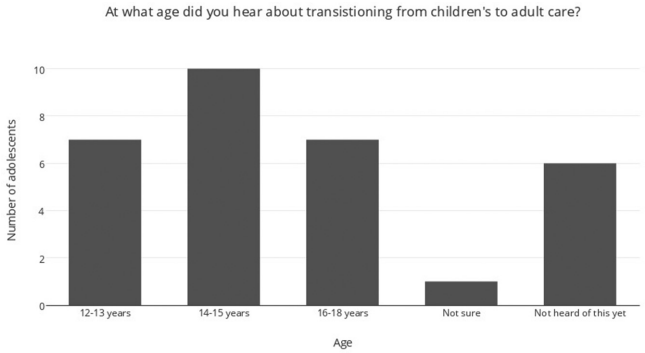
Praise was given for transition services such as a previously organised tour of the adult department, a peer discussion day and an information pack.

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