infections. There was no leukemic transformation or deaths in our study group.

Conclusion Timely and early referral to specialist services will not only prevent morbidity, enable appropriate follow up with early recognition of complications and institution of preventive patient care but also promote family's understanding of and coping with the syndrome.

PS-086 QUESTIONNAIRE STUDY FOR ASSESSMENT OF QUALITY OF LIFE (QOL) IN CHILDREN WITH SHWACHMAN DIAMOND SYNDROME (SDS)

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SDS is a multisystem disorder with heterogenous clinical presentation. Neuro-cognitive impairments, intellectual disability and behavioural difficulties have been reported in previous studies.

Methods and patients We used validated child and parent reporting questionnaires (CHQ-CF87, CHQ-PF50) to explore QoL in children with SDS aged 5–18 years (followed at SDS multidisciplinary clinic at Leeds). 7 children and 1 of their parent completed the self-administered questionnaires. Data was analysed and interpreted as per scoring and interpretation manual provided with the questionnaires. Ethics approval was obtained.

Results Parents scored their child below reference sample for global health and behaviour; they recognised that they worried about their child's health more than other people. There was discrepancy in scoring on some health domains between child and parent CHQ which could represent child's lack of insight or better scoring by parents comparing their child's health and behaviour with their peers.

Qualitative analysis revealed that majority of children reported good health and all experienced mild pain or discomfort. Most were happy, got on with friends, but admitted to arguing and recognised their health/behaviour caused family tension.

Parents reported difficulty concentrating, paying attention and argumentative behaviour. They expressed uncertainty about their child having a healthy life and reported their health/behaviour caused disruption of everyday family activities. Some parents reported their children having difficulties in activities like eating, dressing or going to toilet alone.

Conclusions Our findings identify important qualitative areas of concern and highlight potential areas for providing targeted support to children with SDS and their families.

PS-087 WITHDRAWN

PS-088 TREATMENT OF PRIMARY IMMUNE THROMBOCYTOPENIA BY PREDNISOLONE: 4 MG/KG/J FOR 4 DAYS VERSUS 2 MG/KG/J FOR 6–8 WEEKS

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Background and aims Primary immune thrombocytopenia (ITP) is characterised by accelerated platelet destruction. Corticosteroids have been shown to increase platelet counts. The objective of our study was to compare the treatment with prednisolone at a dose of 4 mg / kg / day for 4 days (Protocol P1) to the treatment with prednisolone at the dose of 2 mg / kg / day for 6–8 weeks (Protocol P2).

Methods We conducted a comparative analytical study between 2 groups: first group (G1) formed by ITP cases followed prospectively between 2010 and 2013. These patients received protocol P1. The second group (G2) collecting the same number of patients followed retrospectively between 1999 and 2008. These patients received protocol P2.

Results Our study included 24 cases: 12 cases in G1 and 12 cases in G2. The response on day three was 25% for G1 and 0% for G2. The response on the fifth day was 66.7% (G1) and 54.5% (G2). The median time to response was 5 days for the 2 groups, with extremes ranging from 3 to 30 days (G1) and 5 to 30 days (G2) (p: 0.09). The complete response occurred in 15 days with a range of 5–90 days for G1 and 30 days with a range of 5–90 days for G2 ($p = 0.04$).

Conclusion Within the limits of our study, we are able to show the interest of high dose of prednisolone during 4 days in the treatment of acute ITP.

PS-089 WHICH DECISION RULES MEET METHODOLOGICAL STANDARDS IN CHILDREN WITH FEBRILE NEUTROPENIA? RESULTS OF A SYSTEMATIC REVIEW AND ANALYSIS

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Background and aim Clinical decision rules (CDRs) have sought to identify the few children with chemotherapy-induced febrile neutropenia (FN) really at risk of severe infection to reduce the invasive procedures and costs for those at low risk. Several reports have shown that most rules do not perform well enough to be clinically useful. Our objective was to analyse the derivation methods and validation procedures of these CDRs.

Methods A systematic review using Medline, Ovid, Refdoc, and the Cochrane Library through December 2012 searched for all CDRs predicting the risk of severe infection and/or complications in children with chemotherapy-induced FN. Their methodological quality was analysed by 17 criteria for deriving and validating a CDR identified in the literature. The criteria published by the Evidence Based Medicine Working Group were applied to the published validations of each CDR to assess their level of evidence.

Results The systematic research identified 612 articles and retained 12 that derived CDRs. Overall the CDRs met a median of 65% of the methodological criteria. The criteria met least often were that the rule made clinical sense, or described the course of action, or that the variables and the CDR were reproducible. Only one CDR, developed in South America, met all methodological criteria and provided the highest level of evidence; unfortunately it was not reproducible in Europe.

Conclusion Only one CDR developed for children with FN met all methodological standards and reached the highest level of evidence.

PS-090 HEREDITARY SPHEROCYTOSIS AND RED CELL INDICES MCHC, MCV, RDW

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Background Hereditary spherocytosis (HS) is a common inherited disorder that is characterised by anaemia, jaundice, and splenomegaly. Clinical severity is variable with most patients having a well-compensated hemolytic anaemia. The primary lesion in HS is loss of membrane surface area, leading to reduced deformability due to defects in the membrane proteins ankyrin, band 3, beta spectrin, alpha spectrin, or protein 4.2. Many isolated mutations have been identified in the genes encoding these membrane proteins; common hereditary spherocytosis-associated mutations have not been identified. The classic laboratory features of HS include minimal or no anaemia, reticulocytosis, an increased mean corpuscular haemoglobin concentration (MCHC), spherocytes on the peripheral blood smear, hyperbilirubinemia, and abnormal results on the osmotic fragility test.
Aim Of the study is to evaluate the role of MCV, MCHC as a screen test to diagnose spherocytosis
Methods In our study are included 60 subjects, 30 children with HS and 30 children-control groups. Our patients with anaemia, jaundice, and splenomegaly are diagnose with HS by incubated osmotic fragility test, performed after incubating RBCs for 18–24 h under sterile conditions at 37°C.
Results We found that 25% of pts. have mild HS, 20% moderate HS, 30% moderate to severe HS and 25% severe HS. In peripheral blood smear 7% of pts. had 0–5 spherocites for field, 30% had 5–10 spherocites for field and 63% had 10–15 spherocites for field. 70% of pts. With HS have MCHC > 38%.

There are a positive correlation between MCHC and spherocites in peripheral blood smear ($r = 0{,}898$, $p < 0{,}001$) and RDW ($r = 0{,}647$, $p < 0.001$), negative correlation between MCHC and MCV ($r = -0{,}437$ $p < 0{,}001$)
Conclusion The dedication of hiperdense erythrociyte today is used as a new tool in diagnosing HS. The determination of MCHC constantly growing with other red cell index, MCV < 80 fl, RDW > 15 obtained from an electronic cell counter usually is enough to suggest for HS.
Key Words Spherocytosis, MCHC, anaemia, children.

PS-091 DISPERSION OF THE QT AND QTC INTERVALS-EARLY MARKER OF ANTHRACYCLINE INDUCED CARDIOTOXICITY IN CHILDREN WITH MALIGNANT HEMOPATHIES

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Background Heart rhythm disorders are one of major adverse effects induced by myocardial anthracicline cardiotoxicity in children with malignancies and that require early diagnosis for effective prevention.
Objectives To investigate the utility of the study of QT and QTc dispersion in children with malignancies treated with anthracyclines.
Methods patients: 40 patients (2–18 years) with malignant hemopathies, treated with anthracyclines. All patients were examined by clinical examen, ECG, Doppler echocardiography, the values of QT dispersion (difference between the maximum and minimum QT interval, manually measure the QT interval, on three successive cardiac cycles) and QT dispersion (Bazett's formula). Dispersion of QT and QTc interval in these patients was compared to similar values from 20 healthy children without cardiovascular history.
Results The increase of QT and QTc dispersion in patients comparative to the control lot, was revealed in 73% cases, usually in those which had a cumulative anthracyclines doses over 400 mg/m^2, with medium values of QTD: 53,33 ± 10,18 msec and QTcD: 66,28 ± 12,8 msec. The increased dispersion of QT and QTc intervals was highlight most frequently in cases with echocardiographycal signs of anthracyclines cardiotoxicity, even only diastolic dysfunction of left ventricle.
Conclusions The significant incidence of increasing the QT and QTc interval dispersion in patients who received treatment with anthracyclines and the correlation with cumulative anthracyclines doses and echocardiographic modifications, especially diastolic dysfunction, proves utility of systematic investigation of QT and QTc intervals dispersion in the full control in the therapy, as an earlier marker for cardiotoxicity of anthracyclines.

PS-092 NUTRITIONAL STATUS OF CHILDREN DIAGNOSED WITH ACUTE LYMPHOBLASTIC LEUKAEMIA AT THE CHILDREN CANCER CENTRE

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Background and aims Acute lymphoblastic leukaemia (ALL) is the most common malignancy among children. Malnutrition remains a major concern for paediatric oncologists. Although studies have shown that malnutrition can negatively affect treatment outcome, results are still controversial. This retrospective cohort study aims at determining the prevalence of malnutrition and its association with treatment outcome and infection among children with ALL treated at the Children Cancer Centre in Lebanon (CCCL).
Methods 108 children and adolescents diagnosed with ALL between April 2002 and May 2010 were enrolled in the study. Anthropometric data were collected from patient's medical record upon diagnosis, at 3 and 6 months, and at the end of treatment. Body mass index (BMI) was calculated for children ≥ 2 years while weight for height ratio was used for patients < 2 years. Patients were considered underweight, stunted, or wasted if their z-scores were <-2SD.
Results The prevalence of malnourished children was 27% at diagnosis and remained almost the same at the end of treatment. The odds ratio of having worse outcome in terms of relapse or death was higher among malnourished children with OR = 2.09, 95% CI = 0.3–13.4 and OR = 1.25 and 95% CI = 0.2–6.9 for death and relapse respectively. However this trend was