

Children's Hospital in Skopje. We explored the characteristics of I.E., together with the causative pathogens, the episodes of febrile neutropenia (FN), the length of antibiotic treatments and the treatments with G-CSF during intensive phases of treatment (Protocol I, M and II).

**Results** From 55 analysed records 24 (43.64%) were male and 31 (56.36%) were female. Mean age at diagnosis was 6.0 years (1.1–15.0). Majority of the patients 43 (78%) were under 10 years and 12 (22%) were over 10 years. All of them experienced 132, 52 and 73 I. E. with 2.4, 0.9, and 1.3 infections per patient during Protocol I, M and II respectively. Regarding to the pathogens 184 (71.5%) were bacterial (102, 30 and 52 in Protocol I, M and II), 45 (17.5%) were viral (20, 14 and 11 in Protocol I, M and II) and 28 (10.8%) were fungal (10, 8, 10 in the three intensive phases respectively). There was a slight predominance of gram positive bacteria in Protocol I [Gram positive 42 (51.85%) versus gram negative 34 (41.97%)], and a very slight predominance of gram negative bacteria in Protocol II [Gram positive 16 (45.71% versus Gram negative 18 (51.42%)]. The infections were treated with antibiotic treatment in average of 23.69, 11 and 15.05 days and the number of treatments with G-CSF were in average 7.22, 2.44 and 9.20 per patient respectively in Protocol I, M and II. The number of episodes of FN in these three phases was 16.4 (29.1%), 4 (7.3%) and 22 (40%).

**Summary/conclusion** Evaluation of the characteristics of I. E. presented that the majority of infectious events were observed in Protocol I and also the length of antibiotic treatment was longer in this phase. But the episodes of FN together with the treatments with G-CSF were higher in Protocol II possible due to the cumulative effect of chemotherapy.

#### PO-0161 TREATMENT OF IRON OVERLOAD WITH DEFERASIROX IN THE PATIENTS WITH THALASSAEMIA MAJOR

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10.1136/archdischild-2014-307384.823

**Background** Iron chelation is an important component of management of transfusion-dependent patients with thalassaemia major. Deferasirox is a relatively new oral iron chelator (US FDA approved in 2005) with the limited experience in children.

**Aims** To present our experience with deferasirox in patients with thalassaemia major (TM) in the context of: effects on serum ferritin level in chronically transfused patients with thalassaemia major, side effects and patients tolerance to the drug and effects on serum creatinin and liver transaminases.

**Methods** Four patients with TM with mean age of 3,1 years (range 2,5–3,5) were included in the study. Mean follow up was 37 months (range 29–42). The disease was diagnosed in early childhood (during the first year of life) with the following signs and symptoms: extreme pallor, jaundice, failure to thrive, poor feeding, irritability, decreased activity and hepatosplenomegaly. Regular blood transfusions were applied to treat chronic hemolytic anaemia.

**Results** The mean serum ferritin (SF) at diagnosis was 471,3 ± 284,4 (range 155–706), and at the start of the treatment with deferasirox 6281 ± 9183,9 (range 767–20000). The number of blood transfusions before the treatment was around 28,3 ± 15,5 units (range 17–46), or about 679,3 ± 608,14 (range 220–1369) ml/kg body weight. Deferasirox was given seven days a week at a dose of 20 mg/kg body weight. The primary outcome variable

was SF level at the start and at the end of the study. Echocardiography was made in all patients and it was normal. MRI-T2\* could not be performed because there was no specific software. The level of SF at the end of the study period was 1862 ± 1312,15 (range 637–3710). Patients were monitored for hepatic and renal toxicity, visual or auditory changes and development of new symptoms. Adverse events were very mild gastrointestinal symptoms in 1 patient and no adverse events in the remaining 3 patients. Elevation of serum creatinin or hepatic transaminases was not observed in any subject. One patient interrupted the therapy as there was a marked fall in SF < 500 ng/L at the end of the therapy. The treatment was well tolerated; suspension of therapy was not required owing to toxicity.

**Summary/conclusion** The results suggest that deferasirox is effective in lowering iron burden, it is well tolerated and has a low potential for toxicity. Long term therapy will be needed to asses the benefits on iron balance and organ damage in chronically transfused patients with thalassaemia major.

#### PO-0162 CLINICAL PROFILE AND OUTCOME OF CHILDREN WITH ANAPLASTIC LARGE CELL LYMPHOMA

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10.1136/archdischild-2014-307384.824

**Background** Anaplastic large cell lymphoma (ALCL) in children is usually ALK positive and is characterised by advanced disease at presentation with a high incidence of extra nodal involvement. We present the clinical profile, treatment and outcome of a small cohort of children with ALCL treated at our centre between January 2004 and June 2013.

**Results** 19 children, 14 boys and 5 girls, aged 18 months to 14 years, with ALCL. 16/19 had lymphadenopathy and 14/19 had fever. Thirteen had high risk, 5 had standard risk and one had low risk disease. 16/19 ALK positive. 17/19 were treated according to the NHL-BFM 90 protocol. Fourteen completed treatment. Three died during treatment; 2 due to infection and 1 had progressive disease. All 3 had high risk disease and were ALK positive. All the three children with ALK negative disease relapsed; one died on relapse treatment, another was lost to follow up a year after completion of relapse therapy and the third with primary cutaneous disease, is alive and well. Of the 11 ALK positive children who completed treatment, only one relapsed and died. The remaining 10 are alive and well with a mean follow up of 17 months (range 8–93 months).

**Conclusion** The majority of our patients are ALK positive and have high risk disease at presentation. 55% of children with high risk disease are alive and well after a mean follow up of 17 months. The overall survival was 65% and event free survival 58% at the time of this analysis.

#### PO-0163 EFFECT OF NUTRITION WITH FOLLOW-UP FORMULA FROM 6TH MONTH OF AGE ON THE COMPLETE BLOOD COUNT AND IRON LEVELS OF INFANTS (PRELIMINARY TRIAL)

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10.1136/archdischild-2014-307384.825

The objective of this study was to compare iron levels of infants fed with supplementary foods because of malnutrition, to those who were supported with complementary foods from sixth months of age, when the breast milk alone is not sufficient. Eighty-four healthy infants aged 6 to 9 months were enrolled. Infants without a nutritional problem, with sufficient iron stores, weighing over 10th percentile were enrolled in the control group, while babies weighing over 3rd percentile, who were not breastfed and did not take a balanced and sufficient diet, without a chronic diseases and who were not born preterm were enrolled in the study group. All of the infants were followed for 3 months. Physical examination findings and complete blood count, iron, iron binding, ferritin, zinc test results were recorded monthly. Infants with malnutrition who could not take breast milk were supported with a follow-up formula (Bebelac). Mean ages were  $7.2 \pm 1.28$  and  $7.3 \pm 1.1$  months in the study and control groups respectively. Statistically significant increase was demonstrated in the MCV and iron levels of the infants supplemented with follow-up formula. MCV decreased significantly in the control group. Mean ferritin values increased from  $53.2 \pm 41.8$  mg/L to  $64.7 \pm 89.7$  mg/L in the study group, while decreased from  $42.8 \pm 34.0$  mg/L to  $34.6 \pm 29.3$  mg/L in the control group. In the study group, significantly higher levels of ferritin were measured in the last assessment, compared to the control group. In conclusion, increase in the iron stores of follow-up formula fed infants was observed during the follow-up period.

#### PO-0164 PAEDIATRIC SOFT TISSUE SARCOMA: A TEN YEAR REVIEW

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10.1136/archdischild-2014-307384.826

**Background and aims** Paediatric soft tissue sarcomas are rare tumours that account for about 7% of all childhood cancers. 50–60% of these are rhabdomyosarcoma (RMS), while the remainder are non rhabdomyosarcoma soft tissue sarcomas (NRSTS). To evaluate the clinical characteristics, treatment modalities and outcome of paediatric soft tissue sarcomas.

**Methods** A retrospective analysis of data from 51 patients diagnosed and treated from 2003 to 2013.

**Results** 51 patients, 30 male and 21 female, median age of 10 years old at diagnosis. 92% showed a mass at diagnosis. The most common site of disease was the extremities (20 cases). Histology: 51% RMS (embryonal: 12 patients; alveolar: 12 patients; other: 2 patients) and 49% NRSTS (PNET 5 patients; malignant peripheral nerve sheath tumour: 3 patients; other: 17 patients). IRS group: 29% group I, 14% group II, 28% group III and 29% group IV. Most patients received multimodality therapy (radiotherapy, chemotherapy and surgery). 31% (16 patients) died due to disease progression: 7 alveolar RMS, 11  $\geq 10$  years old and 13 group III-IV. Time from diagnosis to death was between 7 months and 5 years. Of the 34 living patients, 74% were in first complete remission. The median follow-up time was 38 months.

**Conclusions** Alveolar RMS is more common in the extremities and the embryonal in the genitourinary tract. NRSTS are more common in children  $\geq 10$  years. The absence of metastases and the tumour size  $\leq 5$  cm were associated with a better prognosis ( $p < 0.05$ ).

#### PO-0165 PREDICTORS OF CHRONIC PRIMARY IMMUNE THROMBOCYTOPENIA

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10.1136/archdischild-2014-307384.828

**Background and aims** The primary immune thrombocytopenia (ITP) in children has a favourable evolution in most of cases. Chronic ITP has been noted in 14–30% of the cases. This study reviewed the pattern of presentation and response to therapy in patients with ITP, in order to identify risk factors for chronic disease.

**Methods** We conducted a retrospective study of all cases of ITP in paediatrics and haematological departments of Hedi Chaker University Hospital in Sfax, during a period of 15 years (1995 to 2009). Predictors of chronicity were investigated using Fisher's test with a significance if  $p < 0.05$ .

**Results** 140 cases of ITP were collected including 75 girls (54%) and 65 boys (46%). The average age was 6 years 7 months (3 months to 15 years). The diffuse mucocutaneous purpura was constant and mucosal haemorrhages were present in one third of cases. The mean platelet count was  $25000/\text{mm}^3$  ( $1000/\text{mm}^3$  to  $50000/\text{mm}^3$ ). Therapeutic abstention was the rule in 19 cases (14%), 111 children received corticosteroids (79%) and 10 children received immunoglobulins associated with corticosteroids (7%). Fifteen patients were lost of follow up and evolution has identified two groups: acute ITP in 95 cases (67%), and chronic ITP in 30 cases (21%). The parameters that were found as predictors of chronicity were female sex and age.

**Conclusions** Acute ITP is the most common. Chronic forms are found in one fifth of cases. The determination of risk factors for chronicity can make an early prognosis.

#### PO-0166 THE LANGERHANS CELL HISTIOCYTOSIS IN CHILDREN: STUDY OF 11 CASES

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10.1136/archdischild-2014-307384.829

**Background and aims** Langerhans cell histiocytosis (LCH) is a rare disease of unknown cause with manifestations ranging from isolated granulomatous lesions to life-threatening multi-system organ involvement.

In this study we aimed to evaluate the characteristics, diagnosis, treatment modalities and prognosis of LCH.

**Methods** We conducted a retrospective study of all cases of LCH in paediatrics department of Hedi Chaker University Hospital in Sfax during a period of 16 years (1997–2013). Epidemiologic, clinical, radiological, diagnostic and therapeutic variables were collected.

**Results** We collected 11 cases of LCH. The average age at diagnosis was 3 years 4 months. The patients' presenting symptoms were: exophthalmia (3 cases), polyuropolydipsic syndrome (3 cases), prolonged fever (2 cases), lymphadenopathy (5 cases). Laboratory tests showed diabetes insipidus (3 cases) and bicytopenia (1 case). The diagnosis was confirmed by histopathologic examination in all cases.

Bone was the most frequently affected organ (9 cases) followed by skin (19.2%). Initially, 4 patients had single-system involvement (SS), 3 with multisystem (MS) disease without risk