TREATMENT OF IRON OVERLOAD WITH DEFERASIROX
EFFECT OF NUTRITION WITH FOLLOW-UP FORMULA

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 32, 32 and 73 L.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, 4.4 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 treatment was well tolerated; suspension of therapy was not required owing to toxicity.

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

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 curative 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)

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 hemo lytic anaemia.

Results The mean serum ferritin (SF) at diagnosis was 471.3 ± 284.4 (range 153–706), 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 assess the benefits on iron balance and organ damage in chronically transfused patients with thalassaemia major.
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 ± 1.28 and 7.3 ± 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 ± 41.8 mg/L to 64.7 ± 89.7 mg/L in the study group, while decreased from 42.8 ± 34.0 mg/L to 34.6 ± 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 fed infants was observed during the follow-up period.

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

M Pinheiro, A Maia Ferreira, A Pinto. Pediatrics, Instituto Português de Oncologia, Porto, Portugal

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 ≥ 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 ≥10 years. The absence of metastases and the tumour size ≤5 cm were associated with a better prognosis (p < 0.05).