and receiver operating characteristic (ROC) analysis of most common discriminating indices (Matos & Carvalho, Mentzer Index, RDW Index, Green and King, Ehsani Index) used in differential diagnosis of these two diseases was calculated using MedCalc.v15.2 statistical software. Nonparametric nature of the CBC sample was assessed using the Kolmogorov–Smirnov test. Mann–Whitney test was used to investigate differences between the two groups. Area under the ROC curve was calculated for each index and their differences were assessed. A p -value < 0.05 was considered significant.

Among the 5 tested indices, the Ehsani index correctly diagnosed the highest number of children with β -thalassemia, but failed to properly recognize children with IDA (sensitivity 92%, specificity 46%). The most commonly used Mentzer index showed similar results (sensitivity 88%, specificity 48%). The best ratio between sensitivity and specificity was observed for the new Matos & Cavalho index (sensitivity 74%, specificity 88%) with highest area under the ROC curve. Pairwise comparison of ROC curves observed a significant difference between Matos & Cavalho index and the remaining four tested indices (RDWI $p < 0.0008$; Ehsani $p < 0.0001$; Green and King $p < 0.0001$; Mentzer $p < 0.0001$). Kolmogorov–Smirnov test for normal distribution of CBC values showed a $p > 0.05$ while Mann–Whitney U test for independent samples showed a $p < 0.05$ difference between IDA and β -thalassemia.

Our results show that the most optimal index for discriminating between β -thalassemia and IDA in analysed children is Matos & Cavalho Index.

Therefore, it is more appropriate for discernment than the other analysed indexes. All indexes with low specificity (Mentzer, Ehsani, Green and King) were of low validity as they have a low proportion of IDA correctly identified as such.

295 NON-HODGKIN LYMPHOMA IN CHILDREN: SINGLE CENTER EXPERIENCE DURING 20 YEARS

Ernest Bilić, Zrinko Šalek*, Matej Jelić, Daniel Turudić, Petra Ivančić, Maja Pavlović, Ranka Femenić, Ana Petrović-Gluščić, Lucija Mucavač, Toni Matić, Sara Dejanović Bekić, Josip Konja, Ljubica Rajić. *University Hospital Centre Zagreb*

10.1136/archdischild-2021-europaediatrics.295

Lymphomas are the third most common malignant disease in childhood, after leukemia and brain tumors. The aim of this study is to show stratification by gender and age as well as long term survival in pediatric patients diagnosed with Non-Hodgkin Lymphoma in our center.

Our retrospective analysis included 85 children with newly diagnosed NHL from January 1, 1997 to December 31, 2016. They all have been diagnosed and treated at the Department of Pediatric Hematology and oncology, University Hospital Centre Zagreb.

Out of 85 children with newly diagnosed NHL 48 of them suffered from B-cell NHL ($n = 48$; 56%) while the rest of them had T-cell lymphoblastic lymphoma

(T-LBL) ($n = 20$; 24%) or Anaplastic large-cell lymphoma (ALCL) ($n = 17$; 20%). There were 25 girls and 50 boys (age 3 – 17 years). Overall survival (OS) for the entire group was 78.82%. Diagnose based survival is in the favor of T-LBL – 85.00% in comparison to 81.25% in B-NHL and 64.71% in ALCL.

Our survival rates are not very different from the ones in the other European countries. We expect improved survival

rates after introducing novel treatment that would optimize therapeutic effect and at the same time minimize the risk of severe late toxic effects.

296 ESTIMATION OF THE GLOMERULAR FILTRATION RATE IN CHILDREN WITH HAEMOPHILIA

Zrinko Šalek*, Matej Šapina, Daniel Turudić, Danko Milošević, Ernest Bilić. *Division of Paediatric Haematology and Oncology, Department of Paediatrics, University Hospital Centre Zagreb*

10.1136/archdischild-2021-europaediatrics.296

Haemophilia is rare, inherited and severe bleeding disorder characterised with factor VIII or factor IX deficiency. The estimated glomerular filtration rate (eGFR) is one of the best-performing methods to evaluate kidney function. Glomerular filtration rate cannot be directly measured; however, it can be determined by measuring the clearance of an ideal filtration marker or estimated using predictive formulas. The aim of this preliminary study was to calculate eGFR of paediatric haemophilia patients treated in our centre and assess the correlation of eGFR calculated by creatinine-based and cystatin C-based equations.

In our study, we included 36 boys with moderate or severe haemophilia. Out of a total of 36 patients, 27 had haemophilia A and 9 had haemophilia B.

Their mean age was 11.2 ± 4.31 years, with a range from 3 to 18 years. We investigated the correlation and agreement between two eGFR equations (creatinine-based 'Bedside Schwartz' equation and cystatin-C based equation). Along with applying correlation and linear regression tests, the Bland Altman test was performed to assess the agreement of the results.

Statistically significant differences were found between the mean eGFR values ($p < 0.001$). No significant correlation was found between the two methods ($p = 0.07$). Bland-Altman analysis results showed higher mean eGFR values of bedside Schwartz equation compared to the cystatin-C based formula, meaning that a significant disagreement was found between those two equations. However, within the group of haemophilia B patients, statistically significant positive correlations between the two methods were found, although still a disagreement was observed. Due to the observed disagreements between eGFR within haemophilia patients, further research is needed to find the optimal measure of eGFR. We suggest extending this study on a larger cohort of patients and include other possible eGFR equations.

297 LYMPHOMA OR ALPS?

Cansu Koç*, Gonca Kaçar, Simge Özel Çınar, Suheyla Ocak, Nihan Burtecene, Tiraje Celkan. *Istanbul University-Cerrahpaşa, Cerrahpaşa Medical Faculty*

10.1136/archdischild-2021-europaediatrics.297

ALPS is a rare disease characterized by chronic, non-malignant lymphoproliferation and autoimmunity. The axis of apoptosis is impaired in immunoregulation by mutation in Fas Ligand and Caspase 8 genes.

Lymphadenopathy, hepatosplenomegaly, Direct Coomb's (+), autoimmune hemolytic anemia, ITP are the most

common clinical presentations. Hypergammaglobulinemia is diagnostic of increased CD4-CD8-T cells in peripheral blood. Often it is susceptible to B Cell Lymphoma. In our presentation, we wanted to draw attention to this issue by presenting two different cases, one in the differential diagnosis of lymphoma and the other in the diagnosis of ALPS after long-term lymphoma treatment.

A 6-year-old girl presented in April 2012 with complaints of swelling and night sweats on the left side of the neck. On physical examination, hepatosplenomegaly was absent, and multiple lymphadenopathies were detected in the left cervical chain. No agent was detected in terms of infectious pathologies. There were no pathological cells in bone marrow aspiration performed for malignant diseases, but multiple lymph nodes with cervical, supraclavicular and intra-abdominal hypermetabolic activity were detected in PET imaging. She was diagnosed as Mixed type Hodgkin's Lymphoma by supraclavicular lymph node excision. After ABVD and COPP treatment, radiotherapy was applied to the abdomen, neck and mediastinum. While regression was seen in PET imaging after treatment, the disease was progressed by bone marrow activation and lung parenchymal involvement. Autologous Bone Marrow Transplantation was performed in August 2014, but recurrence was detected for the second time in January 2015. In the follow-up, although the treatment of Brentuximab, Gemcitabine, Paclitaxel and Nivolumab were applied, the disease progressed. Double T Negative Cells 5.3% were detected in immunophenotyping, which was sent for possible immune deficiencies. The history of lymphadenopathy, the predisposition of B lymphoma, the rate of DNT above 2.5%, the presence of a consanguineous marriage between parents and the rate of DNT 11.6% of the sisters who had no complaints, suggested the possibility of ALPS (Autoimmune Lymphoproliferative Syndrome) in the patient. The patient, whose m-Tor inhibitor Sirolimus treatment was started, has been on follow-up for 3 years. Genetic tests of the patient were sent.

A three-year-old male patient was diagnosed as Acute ITP by detecting thrombocytopenia in his examinations with the appearance of bruises on his body after infection. No pathology was observed in bone marrow aspiration.

The response to IVIG and pulse steroid treatments was not good. Due to splenomegaly, coombs positive hemolytic anemia and thrombocytopenia continuing in the follow-up of the patient, DNT cell count was found 7.9% considering ALPS. Genetic tests of the patient were sent. Sirolimus treatment was started for the patient who did not respond well to MMF and oral steroid.

298

OVERVIEW ON THE CURRENT MANAGEMENT GUIDELINES OF IMMUNE THROMBOCYTOPENIA IN CHILDREN

¹Vasile-Eduard Roșu*, ^{1,2}Solange-Tamara Roșu, ^{1,2}Anca-Viorica Ivanov, ^{1,2}Mocanu Adriana-Maria, ^{1,2}Spoială Lia-Elena, ^{1,2}Dușa Cristian-Petru, ^{1,2}Ingrith Miron, ^{1,2}Cristina Gavrilovici. ¹Grigore T. Popa' Medicine and Pharmacy University Iasi, Romania; ²Sfânta Maria' Emergency Clinical Hospital for Children Iasi, Romania

10.1136/archdischild-2021-europaediatrics.298

Our aim is to identify, analyze and compare the international guidelines or national consensus reports on the management of immune thrombocytopenia in children.(ITP).

We performed a systematic search on PubMed database using keywords: 'immune thrombocytopenia in children AND ['children' OR 'pediatric' OR 'paediatric'] AND ['guideline' OR 'consensus'] between 1992 (first guideline) and 2020. We excluded publications written in other languages than English or French and animal studies. A total of 54 papers have been initially found. After exclusion of those that were not relevant or other types of publications than guidelines or consensus (reviews, case series, case reports) we ended up in gathering 44 publications. After full text screening, we excluded papers that did not particularly refer to ITP guidelines but to quality of life, adherence to treatment etc. Finally 6 papers have been found to meet such strict criteria.

They are only six countries in the world published having a specific ITP published in Pub Med: USA with American Society of Oncology Guidelines for Immune Thrombocytopenia, Great Britain, Spain, Italy, Argentina and Japan.

The USA and Italian Guidelines recommend for children newly diagnosed with ITP without bleeding or minor bleeding observation rather than corticosteroids and Immunoglobuline IV (USA). For Children with non-life threatening mucosal bleeding the american guideline suggest corticotherapy no longer than 7 days. For the forms non-responsive at the first line the treatment, the American and Spanish Guidelines indicate thrombopoietin receptors agonist (TPO-RA-Eltrompobag) rather than Rituximab and Splenectomy. According to the spanish guideline corticotherapy is the first choice therapeutic. Generally the primary goals of these guidelines are to review and implement evident based-recommendations. Other treatments include Azathioprine, Cyclophosphamide, Cyclosporine A, Dapsone, Danazol and Myofenolate of Mofetil. In 2018 a joint working group (JWG) of several hematology societies (Germany, Switzerland, Austria) published a European Guideline for adults with ITP but more limitate for children with ITP (no standard treatment for chronic ITP at children).

The splenectomy is universally the last options for treatment in ITP at children.

The general purpose of the Guidelines are the implement new therapies (Eltrompobag/Romiplostim) at children because they are rather than corticotherapy and immunoglobuline iv (which are important side-effects and expansive bugets). At children for Eltrompobag are reported minor or moderate side-effects and no for long term.

299

CHILDHOOD NEUTROPENIA – OUR EXPERIENCE

Nuša Matijašić*, Domagoj Buljan, Izabela Kranjčec, Gordana Jakovljević. *Children's Hospital Zagreb*

10.1136/archdischild-2021-europaediatrics.299

Our goal was to describe demographic characteristics, course and outcome of neutropenic patients.

We used descriptive statistics to demonstrate demographic and clinical characteristics of patients who were referred to pediatric hematologist's examination or hospitalized in the Department of Hematology and Oncology, Children's Hospital Zagreb, due to neutropenia from January 1st, 2009 to December 31st, 2019. Oncology patients with febrile neutropenia were excluded from the study.

Altogether, 100 children were examined or hospitalized in our Department during the 11-year period due to neutropenia, of which 56 were males, with an average age of 3.48