years. The cause of a low absolute neutrophil count (ANC) was autoimmune neutropenia in 37 patients, 31 had chronic idiopathic neutropenia, 27 had infectious neutropenia, 3 had cyclic neutropenia, while 2 were diagnosed with alloimmune neonatal neutropenia. The mean value of ANC at presentation was 732/mm³, and the mean lowest ANC detected during disease course was 600/mm³. Among the 5 diagnostic subgroups, both values were the lowest in patients with cyclic neutropenia in which mean ANC at presentation was 420/mm³ and mean lowest ANC detected during disease course was 180/mm³.

Granulocyte colony-stimulating factor (G-CSF) was given to only 3 patients, whose mean ANC at presentation was 140/mm³. Besides the laboratory finding of persistent severe neutropenia, all 3 patients were prone to recurrent infections. Twenty-three patients had additional cytopenia, out of which 4 had pancytopenia, 13 had anemia and 6 patients had thrombocytopenia. Among patients with additional cytopenia, 5 had positive anti-granulocyte antibodies (21.7%). The mean time to disease resolution was 10.2 months, being the longest in the cyclic neutropenia subgroup, while patients with infectious neutropenia mainly recovered after 21.3 days. The mean follow-up time was 1.6 years.

According to data from our Department, neutropenia is most commonly diagnosed in pre-school children, boys being more frequently affected than girls. More than one third of patients have positive anti-granulocyte antibodies. However, the condition is usually benign, it resolves mainly spontaneously in less than a year, and patients generally do not require G-CSF.

300 IMMUNOTHERAPY IN HIGH RISK NEUROBLASTOMA – TREATMENT ALGORITHM OF COMPLICATIONS

Jasminka Stepan-Giljevic*, Gordana Jakovljevic, Aleksandara Bonevski, Filip Jadrijevic Cvrle, Nada Rajacic, Maja Pavlovic, Izabela Kranjcec. *Department of hematology and oncology „Dr Mladen Cepulic”, Children’s Hospital Zagreb*

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We report our center experience in immunotherapy of high risk neuroblastoma, in particular concerning the most common side effects and their management.

Multidisciplinary team approach will be discussed as well, as it is especially important in care for these patients.

Retrospective review of hospital medical data basis High risk neuroblastoma patients receive intensive multimodal therapy that includes induction, consolidation, and postconsolidation phases. The postconsolidation or maintenance phase implies immunotherapy with anti-GD2 monoclonal antibody dinutuximab beta, applied with the purpose of eliminating any residual tumor cells that may exist after induction and consolidation treatment (multiagent chemotherapy, surgery, high-dose chemotherapy and autologous stem cell transplant).

During the 3-year period (2017 – 2019) 8 high risk neuroblastoma patients received immunotherapy with dinutuximab beta in our center. Patients were in remission confirmed by different methods of investigation. Dinutuximab beta was applied in hospital conditions, as a 10-day continuous infusion, with premedication and concomitant medications that included crystalloid fluids, various pain medications, antihistamines, antiemetics and antipyretics, requiring double lumen

central venous catheter and continuous monitoring of vital functions.

Most of our patients achieved adequate pain control with gabapentin and titrated doses of opioids. Fever was a common side effect easily managed by NSAIDs. In case of allergic reaction the rate of dinutuximab beta was decreased by half and restored to its full rate after resolution of symptoms. In the meantime additional doses of antihistamines, oxygen, salbutamol and racemic epinephrine were applied depending on the clinical condition.

Special care was dedicated to fluid balance supervision due to the risk of capillary leak syndrome. Although 5 of our patients had some degree of capillary leak, only one patient developed significant hypotension which was recognized and treated in timely manner with fluid boluses and epinephrine titrated to effect. Upon resolution of hypotension, the drug infusion was restarted at 50% rate during 2 hours and after that in the absence of recurrent hypotension increased to the full rate.

Postconsolidation immunotherapy with dinutuximab beta has become the standard of care for high-risk neuroblastoma patients. The benefits of this kind of therapy do not come without risks. Having front-line providers who are familiar with immunotherapy and its associated toxicities is critical to the safe and consistent administration of this complex therapy. The team includes physicians of different specialities, nurse practitioners, physician assistants and bedside nurses.

301 DIAGNOSTICS AND TREATMENT OF CHILDHOOD MALIGNANT LIVER TUMORS – A SINGLE CENTER EXPERIENCE

Jasminka Stepan-Giljevic*, Domagoj Buljan, Aleksandara Bonevski, Nada Rajacic, Gordana Jakovljevic, Filip Jadrijevic Cvrle, Maja Pavlovic, Izabela Kranjcec. *Department of hematology and oncology „Dr Mladen Cepulic”, Children’s Hospital Zagreb*

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We report our center experience in medical management of malignant childhood liver tumors Retrospective review of hospital medical data basis In the period of 19 years 13 patients were diagnosed and treated in our institution due to malignant liver tumor (8 males and 5 females). Average age at diagnosis was 4,04 years. 3 of them were born prematurely.

Histopathological type of the tumor corresponded to hepatoblastoma in all cases, except one which is the case of hepatocellular carcinoma. The most frequent PRETEXT group was stage 2. Radiological imaging and biopsy were important in making the correct diagnosis. The usual medical management consisted of neoadjuvant chemotherapy and surgical procedure followed by adjuvant chemotherapy. Overall survival is 76,9% and the average follow-up period 71,1 months.

Malignant childhood liver tumors are challenging to manage necessitating multidisciplinary approach and collaboration.

302 4-MONTH-OLD BOY WITH REFRACTORY THROMBOCYTOPENIA AND WISKOTT-ALDRICH SYNDROME – CASE REPORT

Agnieszka Słowińska*, Karolina Bohdankiewicz, Paulina Dziuba. *Wojewódzki Specjalistyczny Szpital Dziecięcy w Olsztynie*

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The diagnosis of thrombocytopenia in newborns and infants is relatively difficult and it should be diagnosis by exclusion. The most important causes of thrombocytopenia that need to be considered in this group are infections, including sepsis as well as CMV infection, fetal hypoxia, chromosomal abnormalities, bone marrow proliferative diseases and neonatal immune thrombocytopenia.

A rare cause of infant thrombocytopenia is genetically determined, most often X-linked, primary immunodeficiency – Wiskott-Aldrich syndrome (WAS).

Depending on the type of mutation in the WAS gene, encoding the WASP protein, there is a wide spectrum of clinical phenotypes: X-linked thrombocytopenia, X-linked neutropenia and classic WAS. The last one is characterized by a triad of symptoms: thrombocytopenia, usually with small platelets, recurrent infections, and eczema.

This case – report presents the case of a 4-month-old boy with thrombocytopenia resistant to treatment found from the second day of his life, who was diagnosed with full-blown Wiskott – Aldrich syndrome in the following weeks. An additional difficulty in the differential diagnosis in this case was the boy's negative family history and the normal size of the platelets in the tests, which is atypical for WAS. The diagnosis was confirmed by a molecular test which revealed a new mutation, not registered in the ClinVar and HGMD database. The boy's severe course of disease has been brought under control thanks to a bone marrow transplant from his mother in the age of 12 months.

303 CENTRAL VENOUS CATHETER FRAGMENTATION IN BOY WITH HEMOPHILIA A AND INHIBITORS

M Jelić, D Šarić, L Mucavac, S Dejanović-Bekić, M Pavlović, D Turudić, E Bilić. *University Hospital Centre Zagreb*

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Hemophilia A is characterized by deficiency in FVIII and approximately 30% of people with severe hemophilia A are affected by inhibitors.

A 11-year-old boy was diagnosed with severe hemophilia A immediately after birth due to hematoma around right kidney and positive family history. He developed inhibitors on FVIII when he was 12 months of age and therapy was changed in activated prothrombin complex on demand. During next 9 years, recurrent bleeding in the both knees started to occur which was manifested with haemophilic arthropathy.

After several attempt, immune tolerance induction (ITI) was approved by insurance company when he was 10 years old. We started with plasma-derived F VIII in dosage of 100 i.u. per kg BW daily. Titer of inhibitors was negative after 3 months of ITI, and half life of FVIII was normalised after 6 months.

For ease of administering therapy, central venous catheter (CVC) was inserted in right subclavian vein. On the other hand, usage of CVC can be complicated by infections and thrombosis. One year after insertion of CVC, he became febrile and microbiology findings revealed that blood culture was contaminated with *Staphylococcus aureus*, *Acinetobacter ursingii*, *Achromobacter xyloxydans* and *Fusarium proliferatum* which was indication for emergency removal of CVC. Unfortunately, during that surgical intervention, breakage of a

CVC occurred and the tip of the catheter was notably missing.

Emergency MSCT of thorax showed tubular structure located in right ventricle with loop at the entrance of right atrium and descending into the inferior vena cava as well as confluence with hepatic veins. To avoid major surgery, the retained portion was successfully removed by the interventional cardiologist through a femoral vein. During procedure, patient received plasma-derived FVIII to maintain the value of FVIII above 80% and there was no bleeding. Prophylactic antibiotics (meropenem, teicoplanin, liposomal amphotericin B) were continued for 10 days. After three negative blood cultures, negative galactomannan (GM) and 1,3-β-d-glucan (BDG) tests, he was released home and regularly controlled in day hospital. He is now using plasma-derived F VIII 1500 IU every other day as a prophylaxis of hemophilia A and is doing well without bleeding and development of inhibitors.

304 THE CASE PRESENTATION THE DIAMOND BLACKFAN ANAEMIA CAN BE PRESENTED AS THE PARVOVIRUS INFECTION

Liza Collins*, Saadia Rao, Elena Cattaneo, Ramona Onita. *Cambridge University Hospital*

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Background Diamond-Blackfan Anaemia (DBA) is a congenital type of anaemia characterised by pure red cell aplasia and associated with congenital bone abnormalities. It is chronic macrocytic-normocytic anaemia.

DBA is a heterogeneous genetic disease, inherited as an autosomal dominant inheritance in 40 to 45% of cases. The remainder 55 to 60% of cases typically present sporadically.

Case We are introducing the girl who initially was presented with pallor and tachycardia 2 months old. She had Hb 26g/L. Platelets 973 109/L, neutrophils 0.8 109/L, Reticulocyte 9.5 109/L. Blood film showed modest aniso/poikilocytosis, unremarkable WBC, thrombocytosis, no polychromasia.

The flow cytometry performed, was showing:- 65% lymphocytes,- no blast cells. She was transfused, her Hb increased to 66g/L. However she required several blood transfusions due to low Hb in 2 months. She had positive IgG for B19 parvovirus infection. She initially was diagnosed with transitional aplastic anaemia due to B19 infection. However as part of the investigations the elevated level of adenosine deaminase was found. She is currently awaiting the genetic test result, confirming the DBA.

Conclusion It is significantly important to complete the investigation for the DBA if the child presented with aplastic anaemia. Diamond-Blackfan anaemia is a rare disease that carries significant morbidity and mortality if not diagnosed early and managed appropriately.

305 LOW GRADE GLIOMA BRAF V600E INHIBITOR MONOTHERAPY IN A 12-YEAR OLD CHILD

Filip Jadrijevic Cvrle*, Hrvoje Jednacac, Nada Rajacic, Tonci Grmoja, Ana Tripalo Batos, Jasminka Stepan Giljevic. *Department of hematology and oncology „Dr Mladen Cepulic“, Children's Hospital Zagreb*

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