about thrombosis in pediatric cancer population compared to adults. Besides, the role of inherited thrombophilia in all cancer patients with thrombosis is still unclear. The aim of this study was to examine the role of genetic thrombophilic defects in cancer-associated thrombosis in children.

The study included 47 children (13 girls and 34 boys) with newly diagnosed cancer, treated at the Division of Hematology and Oncology, Department of Pediatrics, Clinical Hospital Centre Rijeka, between January 2010 and December 2015. Thirty-six patients had hematological malignancies and 11 patients had solid tumors. The median age was 8.8 years (range 0.4 – 19.3 years). Molecular tests on samples of isolated DNA comprised genetic polymorphisms of Factor V (FV Leiden G1691A), prothrombin (FII G20210A) and the enzyme methylenetetrahydrofolate reductase (MTHFR C677T).

In the study group, 8.5% were heterozygous for Factor V Leiden, 6.4% were heterozygous for prothrombin G20210A mutation, 6.4% were homozygous for MTHFR C677T mutation, and 10.6% had combined thrombophilic defects. The prevalence of thrombophilic defects was in accordance with the prevalence in general population. There was no difference between boys and girls, and between patients with hematological malignancies and solid tumors. The rate of thrombosis was 8.5%. Three patients had upper extremity deep venous thrombosis and 1 had right atrial thrombosis. In half (2/4) of patients combined Factor V Leiden and MTHFR C677T heterozygosity were identified.

Our data suggest that inherited thrombophilia is not related to cancer pathogenesis in children. Inherited thrombophilia may play a role in the pathogenesis of cancer-associated thrombosis. A better understanding of the interactions between genetic and non-genetic causes will help to improve the prophylactic and treatment strategies for thrombosis pediatric oncology patients.

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EXTRACORPOREAL MEMBRANE OXYGENATION AND ALLOGENIC HEMATOPOIETIC STEM CELL TRANSPLANTATION IN A CHILD WITH ACUTE MYELOID LEUKEMIA

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ECMO is rarely used in children with acute leukemia due to possible complications.

We describe a 3.5-year-old boy with acute myeloid leukemia (AML) who required extracorporeal membrane oxygenation (ECMO) support due to severe acute respiratory distress syndrome (ARDS) and septic shock.

At the age of 2 years, he was diagnosed with T-cell lymphoblastic lymphoma and he received treatment according to EURO-LB 02 protocol. At the end of the maintenance therapy leukocytosis with anemia and thrombocytopenia occurred. Bone marrow aspiration showed suspicion of myeloid neoplasm.

Second bone marrow aspiration, which was sent to a foreign hematologic center, confirmed AML. Because of the previous lymphoma which started with eosinophilia, and unusual occurrence of AML early after, immature lymphoid/myeloid

neoplasm with eosinophilia was suspected. AML induction treatment was started, with a plan to proceed to allogeneic hematopoietic stem cell transplantation (HSCT) after chemotherapy, because of poor prognosis. The patient received AIE part of the AML-BFM 2012 protocol without serious complications, but on the 9th day of the HAM part of the protocol septic shock developed. The patient deteriorated and was transferred to the Pediatric Intensive Care Unit. A chest X-ray showed extensive infiltrative shading. *Pseudomonas aeruginosa* was isolated in blood culture. Further development of respiratory failure with ARDS and progression of the septic shock required intubation, mechanical ventilation and inotropes. However, conventional therapy was insufficient so central veno-arterial ECMO was indicated and performed. The ECMO was continued for

22 days. Three days after the ECMO discontinuation he was extubated and respiratory and hemodynamically stable. Bone marrow aspiration after the ECMO treatment showed complete remission so the decision to proceed to HSCT without further chemotherapy was made. Allogeneic HSCT from a matched unrelated donor with myeloablative conditioning was performed 40 days after ECMO discontinuation. Three weeks after HSCT acute skin GVHD grade II responded well to corticosteroid treatment together with cyclosporine. Four weeks after the HSCT steroid-resistant liver GVHD developed but responded well to mini extracorporeal photopheresis (ECP). Five months after HSCT patient is in good condition, with good engraftment, in complete remission, and without GVHD.

In our patient, ECMO support provided survival of septic shock and pneumonia with ARDS, and treatment with allogeneic HSCT provided survival of high-risk leukemia with poor prognosis.

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HEREDITARY SPHEROCYTOSIS ASSOCIATED WITH GILBERT SYNDROME – A CASE REPORT

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Gilbert syndrome (GS) and hereditary spherocytosis (HS) are hereditary diseases with high prevalence rates and similar symptoms. The calculated coexistence of these two diseases is 15-35/million births. Severe hyperbilirubinemia compared with the degree of hemolysis in patients with HS should raise suspicion of associated clinical conditions such as GS, glucose-6-phosphate-dehydrogenase deficiency or thalassemia.

We present the case of a 21-month-old girl with HS and GS. The diagnosis of HS was established by osmotic fragility test results in the age of 40 days.

During infancy the patient had several hemolytic episodes precipitated by upper respiratory tract infections, requiring transfusion therapy. Bilirubin levels were around 100 mmol/l. At the age of 21 months during viral infection, an unusually high unconjugated hyperbilirubinemia was noticed (total bilirubin/direct level 239/13 mmol/l) with mild anemia in the absence of severe hemolysis. As hyperbilirubinemia could not be explained by hemolysis, further evaluation was performed. Genotyping of the conjugation enzyme uridine diphosphate-glucuronosyltransferase confirmed GS. The girl has been followed-up regularly, with consistently higher values of total