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ELIMINATION OF IRON IN HEREDITARY HEMOCHROMATOSIS PATIENTS TREATED WITH ERYTHROCYTAPHERESIS

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Backgrounds. Hereditary hemochromatosis (HH) is an inherited, autosomal recessive disorder of iron metabolism that causes the body to absorb and store an excess amount of iron resulting in the progressive accumulation of iron in the liver, pancreas, heart, joints, and pituitary gland leading to potentially serious complications including cirrhosis of the liver, diabetes, and heart problems. The effective treatment is the regular whole blood removal which causes erythropoesis activation and leads to decrease of iron stores. Red cell apheresis is an optional method for removing of higher amount of erytrocytes in one session. The aim of this study was to evaluate the effectiveness of erythrocytapheresis in the treatment of HH. *Methods.* Repeated erythrocytapheresis were performed in 17 patients with diagnosis of HH (15 x C282Y homozygotes, 2x C282Y + H63D heterozygotes) using Haemonetics MCS 3p cell separator (protocol TAE) in which red cells were removed from patients in 2 - 5 cycles; plasma and buffy-coat were reinfused. Collection time, donor convenience, side effects and red cell yield were recorded and analysed. Samples for hematology and iron studies in patients were drawn, analysed and compared to baseline levels. *Results.* 276 (3 - 70) red cell apheresis in 17 patients (13 male, 4 female), age 49,9 (32 - 67), height 175,7cm (160 - 190), weight 82,8 kg (55 - 110), TBV 5186 mL (3627 - 6501). Procedure time was 32 - 87 min. Mean Hb level decreased from 141,7 g/L (115 - 155) before the procedure to 121,6 g/L (93 - 130). Ferritin values decreased from 1199 ng/mL (268 - 3998) to less than 25 ng/mL (7 - 23,9) in each of patients. The drop in ferritin level was 175 ng/mL (67 - 358) per month and 86 ng/ml (41 - 135) per one apheresis, respectively. *Conclusions.* Procedures were well tolerated by patients, no serious side effects were seen, 21 mild citrate reactions (7,6%) were noted. Red cell apheresis is an effective procedure of iron stores reduction in pacients with the hereditary hemochromatosis. Decrease of iron stores in pacients is individual and depends on many factors.

Funding: Supported by research project MZO 00179906

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THERAPEUTIC LEUKAPHERESIS EXPERIENCE OF A SINGLE CENTRE

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Therapeutic Leukapheresis (TL) is an option in the management of patients with hyperleucocytosis, especially associated with leukostatic symptoms. Nevertheless, its clinical and analytical benefit is not well documented in the literature. The aim of this study was to retrospectively analyse the TL performed in our Centre, between January 1998 and December 2005 and also to evaluate its efficacy and complications. During this period, 28 TL were performed in 15 patients (9 men/6 women), with a median age of 22 years (range 2-78), diagnosed with Acute Lymphoblastic Leukaemia (n=6), Acute Myeloblastic Leukaemia (n=7) and Chronic Myeloid Leukaemia (CML) (n=2). Most of the patients (n=14) initiated TL within one week after the diagnosis. One pediatric patient with CML and an initial white blood cells (WBC) count of 306×10^9/L did not have leukostatic symptoms. The others presented cerebral (lethargy, aphasia, dysarthria, altered vision, intracranial haemorrhage) and/or pulmonary (dry cough, respiratory distress and alveolar haemorrhage) manifestations. Each patient was treated with a median of 2 TL (1-4). Aphaereses were performed in a Cobe Spectra cell separator in the Intensive Care Unit. The mononuclear cells program (MNC) was selected in 20 procedures and the polymorphonuclear cells program (PMN) in the other 8 cases. A median of 3 blood volumes per TL was processed (1-4). An efficacy index (EI) was calculated in order to monitor the procedures: EI = (total collected WBC / total pre-aphaeresis patient WBC) x 100. The median pre-aphaeresis WBC count was 213×10^9/L (65-856), which had a corresponding median leukocrit of 8 ml/dL (2-26). The median EI of all TL was 20% (0-47) and when considering each program, the PMN had a median of 23% (16-47) and the MNC achieved 13% (0-30). The median WBC count, 1 hour and 24 hours after TL, was 174×10^9/L (45-650) and 173×10^9/L (109-491), respectively. Serious complications occurred in 4 patients leading to TL interruption. Those were: respiratory arrest, hypotension, respiratory failure and mucocutaneous haemorrhage; however no deaths occurred. Hypocalcaemia related side effects were observed in 13 patients, but promptly reverted with calcium gluconate administration. After TL, clinical improvement was observed in 6 patients who survived at least for one month. The remaining 9 patients, maintained or worsened their condition and all had an early death (<30 days). The overall survival rate at 6 months after TL was 40%. In summary, in this Centre, the majority of patients who underwent TL were critically ill. Even though, the survival rate was similar to the reported in the literature. The lack of immediate clinical improvement was a sign of a poor prognosis. The PMN program was found to be more effective than the MNC and the EI revealed an easily calculated and reliable indicator. Conclusions were limited due to the reduced number of patients in the study. It is important to find standard indicators to technically and clinically monitor the TL, in order to allow multicentric comparisons from the data available.

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RETROSPECTIVE ASSESSEMENT OF THE GLOBAL QUALITY OF LIFE OF PATIENTS WITH ACUTE MYELOID LEUKEMIA AFTER HSCT FROM NURSES PERSPECTIVES: FINDING FROM A CROSS-SECTIONAL AND RETROSPECTIVE STUDY.

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Backgrounds. The cross-sectional and retrospective study analyses the selected factors which influence global quality of life (QoL) of patients with acute myeloid leukemia (AML) after the hematopoietic stem cell transplantation (HSCT). *Aims.* to verify the applicability of the Czech version of an international generic European Quality of Life Questionnaire - Version EQ-5D for the evaluation of global QoL in patients with acute myeloid leukemia (AML) after HSCT at the Department of Clinical Hematology of the 2nd Internal Clinic in the University Hospital and Medical Faculty of Charles University in Hradec Kralove Czech Republic and to evaluate the global QoL in patients with AML after HSCT at the Department of Clinical Hematology of the 2nd Internal Clinic in the University Hospital and Medical Faculty of Charles University in Hradec Kralove, Czech Republic, 3. to analyse selected demografic, healthly and social factors which influence global QoL in patients with AML after HSCT at the Department of Clinical Hematology of the 2nd Internal Clinic in the University Hospital and Medical Faculty of Charles University in Hradec Kralove, Czech Republic. Patients and Methods. The total number of respondents after the transplantation from 2001 to 2003 was 19 and the return rate of questionnaires was 63% (12 respondents: 9 respondents with AML after autollogous HSCT, 3 respondents with AML after allogenous HSCT. HSCT. The mean age of patients with AML was 47,5 years old (range 27-68) and the male / female ratio was 1,17/1. The Czech version of an international generic EuroQol Questionnaire - Version EQ-5D was used. The influence of monitored factors (age, sex, education, merital status, polymorbidity, nicotinism, religion, type of HSCT and the time lapse from the HSCT) on global quality of life of patients was determined by means of dispersion analysis. Results. The above-mentioned factors proved statistically significant dependence of EQ-5D score and EQ-5D VAS on age (in both cases $p<0,01$), religion (in both cases $p<0,05$), nicotinism (in both cases $p<0,01$), education (in both cases $p<0,05$) and polymorbidity (in both cases $p<0,05$). *Conclusion*: EQ-5D score (dimensions of QoL) and EQ-5D VAS (a subjective health condition) significantly decrease with increasing age, religion, nicotinism, education and polymorbidity on patients with AML after HSCT. The global QoL of patients with AML after HSCT is high (mean EQ-5D score 75,1%, mean EQ-5D VAS 67,5%).

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A 5+5 YEAR EUROPEAN NON-INTERVENTIONAL SAFETY STUDY COMPARING ANAGRELIDE HYDROCHLORIDE (XAGRID) WITH OTHER CYTOREDUCTIVE TREATMENTS IN AT-RISK ESSENTIAL THROMBOCYTHEMIA SUBJECTS

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Backgrounds. Long-term data supporting the use of the different cytoreductive agents in the management of elevated platelet counts remains sparse, particularly when analysing long-term safety. Anagrelide is a selective, non-cytotoxic platelet reducing agent that has been used exten-