

quickly with concomitant HbS removal and ameliorating hyperviscosity. We report about SCD acute painful crisis in a 25-years-old woman, who benefit by automated TREX performed at our Apheresis Service using Fresenius<sup>TM</sup> COM.TEC device with a new dedicated program.

**Table 1. Characteristics of the patient.**

	1 <sup>st</sup> TREX		2 <sup>nd</sup> TREX	
	PRE	POST	PRE	POST
Pain (black, abdominal, legs)	intense	reduced	mild	no
Morphine	i.v. continuously	suspended	no	no
Hct (%) / Hb (g/dL)	26.0/9.2	27.8/9.6	27.7/9.0	30.2/10.5
Hbs (%)	80	28	45	19

**Table 2. Characteristics of eritroexchange.**

	1 <sup>st</sup> TREX	2 <sup>nd</sup> TREX
No./total volume of RBCs* (mL)	8/1955	7/1823
Mean Ht(%) of RBCS* (mL)	56	57
Patient blood volume (mL)	3710	3710
Processed blood volume (mL)	3997	3329
Anticoagulant to the patient (mL)	282	239
Flow rate (mL/min)	37	40
Time of procedure (min)	112	93

RBCs\* = red blood cell units

**Methods.** a blood specimen was drawn in advance to assess compatibility, then cross-matched RBC units were filtered for leukocyte depletion. A detailed informed consent was obtained. Erythroexchange was performed by double-vein technique using a new program (Fresenius HemoCare<sup>TM</sup>, Bad Homburg, Germany) which permits to predict both the final hematocrit and HbS level of the patient. Blood cell count and HbS percentages by current haemoglobin electrophoresis were measured before and after each TREX. The pre-apheresis HbS value permitted to appropriately set the cell separator program with the goal of reducing HbS to less than 30%. Check of post-apheresis HbS allowed verifying the accuracy of instrument predictions. During the procedure the patient was carefully monitored as respect to vital signs (blood pressure, heart rate, oxygen saturation) and occurrence of adverse events, in particular signs of transfusion reaction. Calcium gluconate was administered i.v. to prevent or minimize citrate toxicity. **Results.** we performed two TREX procedures on alternate days; the relevant data are given in the table 1 and 2. Complete clinical remission was obtained with no evidence of alloimmunization or other serious complications. **Conclusions.** our experience confirms the beneficial effects of TREX for SCD pain crisis especially when isovolumetric procedures are carried out with an automated device.

**1484****PREHARVEST PREDICTIONS OF STEM CELL PRODUCTS**

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**Backgrounds.** Collected mobilized peripheral stem cell is the commonly used resource for autologous transplantation. The amount of CD34+ cells in the peripheral blood is used as determinant for starting collection. **Aims.** Aim was to see if not only the amount of CD34+ cells /µl determines the yield of the product but also the percentage of CD34% cells should be considered when to start with collection. **Methods.** 259 aphereses of adult patients suffering from hematological (AML, CLL, NHL, MM) disorders and children with solid tumors (ewing sarcoma, neuroblastoma, rhabdomyosarcoma,) were evaluated. Aphereses were done with the Cobe Spectra<sup>TM</sup>, 3-4 times the blood volume was per-

formed. Measured were WBC, MNC and CD34+Cells in the peripheral blood, amount of CD34+cells /kgBW in the aphereses products, collected CD34+cells/processed liter and efficacy of the procedures (MNCs, CD34+cells.) **Results.** According to the specifications of the transplanting departments (CD34+cells > 2x10<sup>6</sup>/kgBW + back up) 190 (73,35%) of the aphereses were completed successfully. 31 out of 190 (16,31%) successfully completed aphereses were started with CD34+cells<20/µL. In 19 out of 23 aphereses we were successful with CD34+cells <20/µL and >0,2%. **Conclusion:** In collections started with CD34+cells <20/µL the percentage of CD34+ cells is a good predictive factor for successful apheresis, even more so for adults then for children. So don't forget to look at the percentage of CD34+Cells in peripheral blood when you decide to start with stem cell apheresis.

**Table 1.**

CD34+	>0.2%	<0.2%
Adults		
>20/µL	97% (104/107)	76% (23/30)
<20/µL	82% (14/17)	13% (7/53)
Children		
>20/µL	95% (20/21)	92% (12/13)
<20/µL	83% (5/5)	41% (5/12)

**1485****THROMBOTIC THROMBOCYTOPENIC PURPURA POST-HEMATOPOIETIC STEM CELL TRANSPLANTATION**

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Thrombotic thrombocytopenic purpura (TTP) is a rare complication of hematopoietic stem cell transplantation (HSCT): the literature is scant and heterogeneous, little is known about the pathogenesis, except that it appears to differ from that of classical TTP. Plasma exchange (PE) is commonly employed for the therapy, but there are no data that support its use. We present our experience in treatment of two post-HSCT TTPs with PE. From May 2004 to December 2005, 52 patients underwent HSCT, and TTP was diagnosed in 2 of them, respectively on post transplant day 47 and 102. Both patients received HSCT from HLA-compatible related donors. TTP was defined as the simultaneous occurrence of red cell fragmentation, laboratory findings of haemolysis with negative direct and indirect antiglobulin test, high LDH level, red cell transfusion requirement and thrombocytopenia caused by consumption, in the absence of disseminated intravascular coagulation. PEs were performed using fresh frozen plasma as replacement fluid. PE was well tolerated, but the two patients had no response to the treatment. One patient died because of fungal infections. Our experience confirm the data of the recent literature. TTP is a rare and serious complication of hematopoietic stem cell transplantation and further, systematic studies are necessary for a better knowledge of its incidence, treatment and outcome.

**1486****QUALITY OF LIFE OF ONCOLOGICAL AND HEMATOONCOLOGICAL PATIENTS AFTER THE HSCT: FINDING FROM CROSS-SECTIONAL AND RETROSPECTIVE STUDY.**

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**Backgrounds.** The cross-sectional, retrospective and descriptive study evaluates quality of life (QoL) of patients after the hematopoietic stem cell transplantation (HSCT) at the Department of Clinical Hematology of the 2nd Internal Clinic of the University Hospital and Medical Faculty of Charles University in Hradec Kralove, Czech Republic from 2001 to 2003. **Aims.** 1. to verify the applicability of the Czech version of an international generic European Quality of Life Questionnaire - Version EQ-5D for the evaluation of QoL in patients after the HSCT at the Department of Clinical Hematology of the Second Internal Clinic in the University Hospital and Medical Faculty of Charles University in Hradec

Kralove, Czech Republic, 2. to evaluate the QoL of patients 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 3. to analyse influence of selected factors on QoL of patients after the HSCT at the Department of Clinical Hematology of the Second 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 was 95 and the return rate of questionnaires was 72.1% (71 respondents - 39 men and 32 women) and we could evaluate 100% of them. Their average age was 55.5 years. We used the Czech version of an international generic European Quality of Life Questionnaire - Version EQ-5D. The influence of monitored factors (type of transplantation - autologous, allogeneous, age, sex, education, polymorbidity, marital status, religion and the time lapse from the HSCT) on the QoL of patients after the HSCT was determined by means of dispersion analysis. *Results.* The above-mentioned factors proved statistically significant dependence of EQ-5D score and EQ-5 VAS on age (in both cases  $p < 0.01$ ), polymorbidity (in both cases  $p < 0.01$ ) and on religion (in both cases  $p < 0.01$ ). The influence of other factors on EQ-5D score and EQ-5D VAS was not proven as statistically significant. Conclusion: EQ-5D score (dimensions of QoL) and EQ-5D VAS (a subjective health condition) significantly decrease with increasing age and with a higher number of associated diseases. They are significantly higher in patients who believe in God compared to patient without religious beliefs. Based on our study we can further state that the QoL of our patients after the HSCT at the Department of Clinical Hematology of the Second Internal Clinic of the University Hospital and Medical Faculty of Charles University in Hradec Kralove, Czech Republic, is very high, which is seen from mean EQ-5D score (72.5%) and mean EQ-5D VAS (76.5%) values.

**1487****QUALITY OF LIFE OF LITHUANIAN CHILDREN SUFFERING FROM CANCER**J. Makari,<sup>1</sup> A. Zaborskis<sup>2</sup><sup>1</sup>Kaunas University of Medicine Hospital, KAUNAS, Lithuania; <sup>2</sup>KMU Institute for Biomedical Research, KAUNAS, Lithuania

Cancer is the most often cause of death in children. According to data of the Lithuanian cancer registry, in the last decade 70'100 new cases of children cancer were diagnosed yearly. In Lithuania, the quality of life of children suffering from cancer until now has not yet been evaluated properly. Aims. The aim of the study is to increase the understanding of the quality of life of Lithuanian children suffering from cancer. Methods. The study started in February of 2005 in the Division of Oncology and Hematology at the Clinical Hospital of Kaunas University of Medicine and in the Division of Oncology and Hematology at the Vilnius University Children's Hospital. During one year, 63 children aged 2'18-year and their families were invited to participate in the study. In the sample, 55% of children suffered from hematoblastosis, 13% from CNS tumors, and 32% from solid tumors of other localizations. The children and their parents were questioned within 2 to 6 weeks from the date of diagnosis. We used the PedsQL (Pediatric Quality of Life Inventory TM) questionnaire initially developed to evaluate the quality of life of children between the ages of 2 and 18. The questionnaire was translated from the original English version (designed by Dr. James Varni and the Mapi Research Institute) into Lithuanian according to the linguistic validation criteria. Children aged 5'7 were interviewed by researchers while older children and parents of children from all age groups filled out the questionnaires by themselves. *Results.* 36.1% of children aged 8'18 stated that they had low energy often or almost always; 54.9% of their parents thought similarly. Children aged 5'7 less often complained of having low energy when compared with older children; none of the children in this age group complained of always being tired. 40.0% of parents whose children were 2'4-year-old felt that children often or almost always needed more energy to play. Among 8'18-year-olds, 27.8% of respondents stated they never felt scared and sad; 22.2% of the respondents did not feel angry because of their present disease. In this age group, 9.7% and 16.1% of parents felt scared, sad and angry respectively. 33.3% of respondents stated that they sometimes felt uneasy that their disease will relapse; parents worried about this more often - 41.9%. Furthermore, in the 8'18-year age group, 27.8% of children stated feeling pain often or almost always, whereas this complaint was stated by 38.8% of their parents. Among 5'7-year olds, often or almost always felt pain was reported by 12.0% of children, whereas the child's pain was indicated by 28.9% of parents. Among 2'4-year-old children, 46.7% of parents stated that their children often felt or almost always felt pain. Conclusions. Children evaluated their quality of life as being better when

compared with their parents. Younger children evaluated their quality of life as better than older children.

**1488****THROMBOPHILIC MUTATIONS IN THALASSEMIA AND  $\beta$ (0)-THALASSEMIA**F. Kouskou,<sup>1</sup> L. Benetatos,<sup>2</sup> V. Alymara,<sup>2</sup> A. Bassou,<sup>2</sup> K.L. Bourantas<sup>2</sup><sup>1</sup>University Hospital of Ioannina, IOANNINA, Greece; <sup>2</sup>Dept. of Haematology, IOANNINA, Greece

*Backgrounds.* The thalassaemic patients present a higher than normal incidence of thromboembolic events including cerebral thrombosis, deep venous thrombosis and pulmonary embolism. The study of the hemostasis of thalassaemic patients revealed increased circulating platelet aggregates, a significant shortening of platelet life span, increased concentrations of urinary metabolites of thromboxane A2 and prostacyclin, low levels of protein C and protein S. On the other hand, a number of mutations are associated with an increased risk of thrombosis. Heterozygotes for the Factor V G1691A (Leiden) mutation experience 2-5 times the normal risk of thrombosis, while homozygotes' risk is 80 times the risk in non carriers. The MTHFR G677T and A1298C mutations are associated with homocysteinemia and increased risk of cerebrovascular disease and peripheral artery disease. Other thrombophilic mutations are the Prothrombin G20210A, FV H1299R (R2), Factor XIII V34L,  $\beta$ -Fibrinogen '455 G-A, PAI-1 4G-5G, GPIIIa L33P (HPA-1), ACE I/D, Apo B R3500Q and Apo E2/E3/E4. *Aims.* The aim of the present study was to check whether the presence of a thrombophilic mutation in a thalassaemic patient increases the risk for the development of a thromboembolic event.

**Table 1. Number and percentage of thrombophilic mutations.**

Mutation	Thalassaemic (20)			Non Thalassaemic (20)		
	Total	Heterozygous	Homozygous	Total	Heterozygous	Homozygous
Factor V G1691A (Leiden)	20			20	3 (15%)	
Factor V H1299 (R2)	20	3 (15%)		20		
Prothrombin G20210A	20	3 (15%)		20	4 (20%)	
Factor XIII V34L	20	11 (55%)		20	5 (25%)	1 (5%)
$\beta$ -Fibrinogen '455 G-A	20	11 (55%)	1 (5%)	20	8 (40%)	
PAI-1 4G/5G	20	15 (75%)	4 (20%)	20	10 (50%)	6 (30%)
GPIIIa L33P (HPA-1)	20	1 (5%)		20	5 (25%)	
MTHFR C677T	20	8 (40%)		20	9 (45%)	1 (5%)
MTHFR A1298C	20	7 (35%)	4 (20%)	20	13 (65%)	1 (5%)

*Methods.* We have screened an unselected group of 20 patients, 17 men and 3 women. 16 of them had  $\beta$ -thalassaemia major, 2 intermediate  $\beta$ -thalassaemia and 2 S- $\beta$ (0)-thalassaemia. The mean age of the patients was 30,5 years. One of the patients presented ulcer of the lateral malleolus, another had avascular necrosis of the femoral head. A group of 20 sex- and age-matched healthy individuals served as control group. DNA analysis was performed by polymerase chain reaction and reverse hybridization. Both patients and healthy individuals were checked for 9 mutations: FV G1691A (Leiden), FV H1299R (R2), Prothrombin G20210A, Factor XIII V34L,  $\beta$ -Fibrinogen '455 G-A, PAI-1 4G/5G, GPIIIa L33P (HPA-1), MTHFR C677T, MTHFR A1298C. *Results.* Table 1 shows the number and the percentage of heterozygotes and homozygotes of the mutations that were studied. The profiles of the mutations of the two thalassaemic patients that were having clinical manifestations of hypercoagulability were the following: the 34-year old man with