

16mg/kg/day. The target area under the curve (AUC) was defined as 17.500 $\mu\text{g}\cdot\text{h}/\text{l}$ and was assessed on day 1 and day 4 based on 3 blood samples. Dose adjustment was performed before the second dose. Primary endpoint was A&E, secondary endpoints were veno-occlusive disease (VOD), graft versus host disease (GvHD). A risk factor analyses was performed using logistic regression.

Results: Between 6/2003 and 6/2006 60 pts were included (30 IVdtBU and 30 POBU). The IVdtBU and POBU groups were comparable regarding age, sexe, indication for SCT, match-grade and donor-source. The A&E rates for the IVdtBU and POBU were 83.3% and 40%, respectively (OR 7.3; 95% CI 2,1-26; $p=0.002$). VOD occurred in 33% and 13% of patients, respectively ($P=0,1$). No difference in GvHD (\geq grade2) was found. Average AUC after the first dose was 20.710 $\mu\text{g}\cdot\text{h}/\text{l}$ and decreased after targeting to 18.920 $\mu\text{g}\cdot\text{h}/\text{l}$. However, interpatient variation remained comparable. The pharmacokinetic data showed a large difference between the clearances of busulfan in children as compared to adults. Clearance correlated well to body surface area and seemed to be constant at all ages, whereas clearance divided to bodyweight decreased as a function of age.

Conclusions: IVdtBU resulted in higher A&E rates compared to POBU. A non-significantly trend to more VOD in the IVdtBU group was observed. Because of the generally high AUC of day 1 starting dose might be lowered. Once daily dosing of IV busulfan with TDM was feasible in routine clinical practice and improved the A&E rates for SCT in children.

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OUTCOMES OF CORD BLOOD TRANSPLANTATION FOR HURLER'S SYNDROME. AN EUROCORD-WORKING PARTY INBORN ERRORS EBMT SURVEY

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Background: Hurler's syndrome (HS), the most severe form of mucopolysaccharidosis type-I causes progressive deterioration of the central nervous system and death in childhood. Allogeneic-stem cell transplantation (SCT) before the age of two years halts disease progression and prolongs life. Graft-failure and mixed chimerism limits the success of SCT for HS. (Unrelated) cord blood grafts are suggested to be a good alternative source for bone marrow.

Methods: Patient registered to EUROCORD-database and EBMT-Hurler syndrome database were included. Between 1991 and 2004 forty-two patients received an (unrelated)-cord blood transplant. The children were evaluated for engraftment, adverse event and effects on disease symptoms. A risk factor analyses was performed using logistic regression.

Results: From 40 patients enough data was available. After a median follow up period of 14 (7-84) mths the "alive and engrafted" rate was 69% after first SCT. Donors used were: 4 HLA-identical family and 36 unrelated cord blood (23 mismatched: 18 5/6-matched and five 4/6-matched). With the exception of 2, all patients received a myeloablative conditioning regimen. Cell dose used was median 7.9 (1.5-32) $\times 10^7$ NC/kg and 2,6 (0,7-25) $\times 10^5$ CD34+/Kg. In multivariate logistic regression analyses, year of transplantation (≤ 2000 vs. ≥ 2001 : A&E 40 vs 84%: OR 7,5 range 1,7-30, $p=0.007$) and NC-dose of less than 5×10^7 NC/kg (OR 6,4, range 1,7-39: $p=0.045$) were found to be risk factors for graft-failure. Only 2 of the 27 patients A&E patients showed a mixed chimerism (86 and 92%: still increasing). All patients had normal enzyme levels. Acute-GvHD (grade >1) was observed in 15%, while chronic-GvHD was seen in 19% (extensive 8%) of patients at risk. SCT improved somatic features of HS.

Conclusion: Outcome following cord blood SCT for Hurler's syndrome is encouraging. (U)CB is a good alternative stem cell

source and might even be preferential since cord-blood appeared to increase the likelihood of sustained engraftment resulting in full-donor chimerism and normal enzyme levels.

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CLINICAL OUTCOME OF HUMAN HERPES-6 REACTIVATION AFTER HAEMATOPOIETIC STEM CELL TRANSPLANTATION IN CHILDREN

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Background: Although human herpes virus 6 (HHV6) is known to reactivate during haematopoietic cell transplantation (HCT) and is suggested to be associated with severe clinical manifestations in adults, the clinical significance in children remains controversial. In this study we investigated the incidence of HHV6 reactivation and HCT-associated morbidity and mortality in children.

Methods: Between 1/2004 and 4/2006 59 patients, median age 6.6y (0.1-18.1), underwent 68 allogeneic HCTs. By quantitative PCR HHV6, EBV, CMV and adenovirus (AdV)-plasma loads were measured once a week. Clinical features, engraftment, number of transfusions, HCT-associated mortality and morbidity (like GvHD) were monitored. Antiviral treatment for HHV-6 reactivation was only given from 4/2005 for those with clinical symptoms assumed to be associated with HHV6. HHV6 reactivations were grouped in group I (no HHV6), group II (loads $<1000\text{cp}/\text{mL}$) and group III (loads $>1000\text{cp}/\text{mL}$). CMV, EBV and AdV-reactivations were treated according to local guidelines. A risk factor analyses was performed using logistic regression.

Results: 36 HLA-id and 23 HLA non-id grafts were used: 44 bone marrow/PBSCs and 15 cord blood grafts. Median follow up was 17 (5-35)mths. HHV6 reactivation occurred in 40/59 (67%) with 33/40 (82%) occurring within the first 30 d post-HCT. 23/59 (39%) had HHV6 loads above 1000cp/mL (group III). Groups did not differ regarding sex, age, donor source or HLA-disparity. In multivariate analysis HHV6 reactivation was associated with a lower survival in group III (OR 0,16; range 0,03-0,89; $p=0.035$) as well as with more multiple viral reactivations ($p=0,049$: OR 5,5; range 1,1-29) were seen in this group. In 11/13 multiple viral reactivation HHV6 was the first viral reactivation. There was a non-significant trend for more grade 2-4 acute-GvHD ($p=0,058$ OR 4,3; range 0,8-18) in group III. HHV6 didnot influence period of neutro-trombocytopenia.

Conclusion: HHV6 reactivation is common after HCT in children and is associated with a higher rate of multiple viral reactivations, aGvHD and with a lower survival rate. Although the exact role of HHV6 reactivation in transplantation associated morbidity and mortality has to be elucidated, early detection and initiation of therapy might influence the outcome.

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JOB SATISFACTION AMONG ADVANCED PRACTICE NURSES WITHIN THE PEDIATRIC BLOOD AND MARROW TRANSPLANT CONSORTIUM

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Introduction:

Advanced Practice Stem Cell Transplant (SCT) Nursing provides unique challenges associated with the medical science of SCT, current healthcare environment and intense relationships with patients and families due to the demanding nature of this therapy. Although job satisfaction among nurses has been discussed in the literature, the focus has been on work related and psychological stresses of SCT staff nurses working in inpatient SCT units^{1,2}. To date little if any research has been done exploring job satisfaction specific to Advanced Practice SCT Nurses (APSCTN) who have become integral members of the pediatric SCT team.

Methods:

Pediatric Blood and Marrow Transplant Consortium (PBMTCC) Nursing Discipline members completed a simple questionnaire