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serotherapy alone, 6 pts with reduced intensity regimens, and 3 pts received no conditioning. Median age at the second SCT was 6.2 years (range, 0.3–14.2 years) and the median time from the first SCT to the second SCT was 10.3 months (range, 1.2–84 months).

Results: 40 patients had initial engraftment, median time to ANC $\geq 500 \times 10^6$/l was 19 days (range, 9–28 days), and median time to a platelet count $\geq 20 \times 10^9$/l was 26.5 days (range, 10–100 days). Acute grade ≥ 3 GVHD, developed in 7 pts (13.7%), mild VOD of the liver developed in 2 pts (3.9%).The 10-year overall (OS) and event free survival (EFS) were 66% and 50% respectively. When survival was evaluated by the interval between the 1st and 2nd SCT: < 6 months (19 pts) and ≥ 6 months (32 pts), the 10-year OS was 55% and 73% for the 2 cohorts respectively ($P = 0.05$), and the 10-year EFS was 27% and 67% for the 2 cohorts respectively ($P = 0.005$). With intervals of < 12 months (26 pts) and ≥ 12 months (25 pts), the 10-year OS was 64% and 70% for the 2 cohorts respectively ($P = 0.4$), and the 10-year EFS was 42% and 63% for the 2 cohorts respectively ($P = 0.2$). When survival was evaluated by underlying disease, the 10-year OS was 59%, 75% and 59% for pts with NMHD, IMD and leukemia respectively ($P = 0.3$), and the 10-year EFS was 55%, 50%, and 59% ($P = 0.5$) for the same groups respectively.

Conclusions: Second SCT in children is relatively well tolerated and is associated with reasonable survival. OS and EFS appear to be considerably better when the interval between the 2 transplants is $\geq$ 6 months.

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RESULTS OF HEMATOPOIETIC STEM CELL TRANSPLANTATION IN PATIENTS WITH LEUKOCYTE ADHESION DEFICIENCY TYPE I

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Objective: Hematopoietic stem cell transplantation (HSCT) is the only curative treatment of some congenital immunodeficiency diseases especially leukocyte adhesion deficiency type-I (LAD-I).

Methods: Six patients (3 girls, 3 boys), received allogeneic HSCT between 2007 and 2008. One patient received stem cell from her healthy full human leukocyte antigen (HLA) matched sibling, three patients from their full matched other related (one from father, one from mother and one from uncle which verified with high resolution HLA matching method), one patient from his mother (haploidentical) and one patient from his father with one antigen mismatch. The graft source was bone marrow in 4 patients and peripheral blood stem cells (PBSC) in others. Median age at HSCT was 17 months (10.2–87.8 months). All patients were conditioned with Fludarabine, Melfalan and Antithymocyte globulin (ATG). For graft-versus-host disease (GVHD) prophylaxis, we used Cyclosporine and Methylprednisolone.

Results: All patients engrafted. Median time of Absolute Neutrophil Count $\geq 0.5\times10^9$ /L was +13 and median time of Platelet recovery $\geq 20\times10^9$ /L was +16. Median follow-up of patients was 8 months (range, 1–20 months) and at present five (83%) patients are alive with sustained engraftment and without recurrent infection and improvement of their failure to trive. Three patients developed acute GVHD after transplantation that one of them died because of acute GVHD one month after transplantation. No chronic GVHD occurred. Full chimerism was achieved in one patient and mixed chimerism was achieved in four patients.

Conclusions: Early diagnosis and HSCT of LAD-I rescue patients from lethal disease but not all of patients have full match siblings and others must transplanted from other related, haploidentical relative, non relative full match or cord blood. Analysis of our patients who developed mixed chimerism suggested that this mixed chimeric state resulted in a reversal of the LAD-I phenotype.

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STABLE MIXED CHIMERISM AFTER NON-MYELOABLATIVE TRANSPLANT FOR SICKLE CELL DISEASE: NORMAL HEMATOPOIESIS DOMINATES OVER ABNORMAL IN MIXED CHIMERISM

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Allosensitization due to transfusion therapy has been a barrier to engraftment in patients with sickle cell (SC) disease, requiring the use of more intense conditioning. In a mouse model, sensitization to bone marrow cells (BMC) results predominantly from the humoral immune response. We added alemtuzumab (anti-CD52) lymphodepletion for conditioning recipients for a hematopoietic cell transplant. 2 cycles of alemtuzumab were administered with the rationale of eliminating memory B cells by inducing them to undergo homeostatic proliferation after the 1st round of depletion. Conditioning consisted of alemtuzumab (10 mg/day, days -53— -50 and days -24 —-21), fludarabine (30 mg/m^2/day days -5 —-3), 200 cGy TBI (day -1), matched related donor BM infusion day 0, followed by cyclosporine and mycophenolate mofetil (day-1—>10 months). 2 patients have been transplanted. Patient 1 was 7 at transplant, with a history of multiple pain crises, episodes of acute chest syndrome, and extensively treated with exchange transfusion therapy and hydroxyurea. She received 14.5×10^6kg CD34 cells from her HLA-identical sibling donor with SC trait, showing 78% donor engraftment day 30. Her RBC production remained phenotypically trait which is compatible with donor hematopoiesis > 615 days post-transplant, with donor lymphoid and myeloid chimerism consistently 20–30%. Immunosuppression was discontinued 10 months post-stem cell transplantation (SCT). Patient 2 was 9 at transplant with multiple pain crises, 2 episodes of acute chest syndrome before transplant, and had been treated with exchange transfusions for 7 years. He received HLA-matched sibling donor BM, processed to deplete T- and B cells. The graft contained 5.24×10^6/kg CD34, 0.55×10^6/kg αβ-TCR and 0.35×10^6/kg FC cells. He has been transfusion-independent post-SCT, with 100% donor RBC production and chimerism levels at 20–30% donor by FISH >851 days post-transplant. Immunosuppression was discontinued at 23 months post-SCT. Neither patient had GVHD, transplant-related toxicity, or SC complications since transplant. Data suggest that the addition of an alemtuzumab split-course to a nonmyeloablative transplant regimen allows hematopoietic cell engraftment of allosensitized recipients with SCD without GVHD. Stable mixed chimerism of myeloid and lymphoid lineages is accompanied by full donor erythropoiesis. Stable mixed chimerism may also have been achieved due to prolonged post-SCT immunosuppression from MMF.

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BONE MINERAL DENSITY, LEAN BODY MASS, CALCIUM AND VITAMIN D INTAKE IN CHILDREN AND ADOLESCENTS AFTER HEMATOPOIETIC STEM CELL TRANSPLANTATION

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Introduction: Hematopoietic stem cell transplantation (HSCT) has been associated with decreased food intake, malnutrition, reduced lean body mass and reduced bone mineral density (BMD), which might result in increased morbidity and mortality. The purpose of this study was to evaluate the BMD, lean body mass, calcium and vitamin D intake in children and adolescents before and 6 months after HCT.

Patients and Methods: Forty patients (25 boys and 15 girls; mean age: 10,1 ± 4,2 ± yrs) who were consecutively submitted to allogenic HSCT between September 2006 and March 2008 and survived after 6 months were included in the analysis. BMD was evaluated by DXA (HOLOGIC-1000) at the lumbar spine (LS) and whole body (WB), lean body mass by bioelectrical impedance analysis and food intake by a 24 hours recall and food frequency questionnaire.

Results: LS BMD was 0,673 ± 0,167 g/cm^2 in the first evaluation and 0,662 ± 0,78 g/cm^2 in the second evaluation ($p > 0.05$). The WB BMD was 0,850 ± 0,164g/cm^2 in the first evaluation, reducing to