statistically significant difference (p 0.02) compared with those subjected to HT as a first choice.
OS of 70% was reported at 40 months. OS and DFS for children with leukemia was 75% and 40%, respectively. For children with immunodeficiencies it was 65% and 15%, respectively, with a 40-month follow-up

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Factors Associated with CMV Disease in Pediatric Hematopoietic Stem Cell Transplantation

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Purpose: Cytomegalovirus (CMV) infection remains a significant source of morbidity in pediatric hematopoietic stem cell transplantation (HSCT). Current strategies for mitigating the effect of CMV on outcome include weekly measurement of CMV viral load in the blood through post-transplant day 100 and use of acyclovir prophylaxis in the peritransplant period in patients at risk for CMV infection (patients in which the HSCT donor or recipient had CMV IgG seropositivity indicative of latent infection). Risk factors predicting CMV infection in pediatric HSCT are not well understood, and factors affecting recurrence of CMV viremia following an initial episode have not been reported.
Methods: We performed a retrospective review of consecutive cases at our institution between 2011 and 2014 where the recipient was at risk for CMV infection. We calculated odds ratios (OR) and 95% confidence intervals (CI) of CMV reactivation as a function of HSCT characteristics. This study was approved by the Institutional Review Board of the Dana-Farber Cancer Institute.
Results: Out of a total of 91 at risk patients, 26 (29%) patients had CMV infection (defined as CMV viremia without target organ involvement) occurring at a median of 46 days following HSCT (range: 9-127). One patient died from biopsy-proven CMV pneumonitis. There was a trend towards recipients with underlying malignant conditions having increased risk of CMV infection compared to others (OR=2.3; 95% CI=0.9-6.0; p=0.08). There was a significantly increased risk of CMV infection in recipients of an umbilical cord blood compared to other sources (OR=9.45; 95% CI=1.8-50.6; p=0.009). Patients with acute graft-versus-host disease (aGVHD) had a significantly increased risk of CMV infection (OR=3.6; 95% CI=1.1-12.1; p=0.04). All patients with viremia received a 14-day course of antiviral and immunoglobulin therapy. Patients who failed to clear the virus completely from the blood at the end of 14 days of therapy had no increased risk of CMV recurrence (p=0.2). A total of 6/26 (23%) HSCT recipients experienced CMV recurrence, at a median of 33 days (range 9-74) following initial CMV clearance. In the subset of recipients who experienced a recurrence of CMV infection, there was a trend toward increased treatment-related mortality (TRM; OR=9.5; 95% CI = 0.7-132; p=0.09). However, among all recipients at risk, CMV viremia was not associated with increased TRM (p=0.7).
Conclusions: Pediatric patients seropositive for CMV who receive umbilical cord HSCTs have increased risk for CMV viremia in the post-HSCT period. CMV infection is associated with aGVHD. If treated with antivirals, recurrence of CMV can be prevented. Multiple episodes of CMV viremia may be associated with increased TRM. This study broadens understanding of CMV disease in pediatric HSCT, and is the first to analyze factors influencing recurrence of CMV infection.

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Late Onset Pulmonary Arterial Hypertension after Successful HSCT for Familial HLH

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Familial hemophagocytic lymphohistiocytosis (FHLH) has often been associated with high transplant-related mortality, typically from a variety of causes in the first 100 days post-HSCT. We report a case of occult pulmonary hypertension that developed 3 years after HSCT for FHLH. The patient was born at 28 weeks with a NICU course consisting of mild respiratory distress syndrome and unilateral grade II intraventricular hemorrhage. At 2.5 months of age, he developed fevers, splenomegaly, progressive pancytopenia, hypofibrinogenemia, hyperferritinemia, and bone marrow hemophagocytosis. He underwent initial treatment per HLH-2004 with good response. Genetic analysis revealed homozygous perforin 1 mutations. He proceeded with a 5/6 cord blood transplant with Bu/Cy/VP-16/ATG conditioning and GVHD prophylaxis with cyclosporine and steroids. He achieved successful engraftment with stable 100% donor chimerism.
Post-HSCT course was complicated by engraftment syndrome, steroid-responsive grade II skin and gut aGVHD, severe VOD treated with defibrotide, and prolonged intubation. He developed persistent renal failure, presumably due to chronic calcineurin toxicity. He underwent cadaveric renal transplant 10 months after HSCT. Clinical care over the next two years was all outpatient, but complicated by BK nephropathy.
Two months prior to his death, he presented with tachypnea and oxygen desaturation. CXR showed mild peribronchial opacities. Respiratory viral panel and echocardiogram were normal. His symptoms resolved quickly with albuterol. He was re-admitted a week later with similar symptoms. Repeat CXR and echocardiogram were unchanged. Due to persistent symptoms, he underwent bronchoscopy that was positive for *P. jirovecii* and was treated with high-dose Bactrim. One month after completing treatment, he presented to an outside ER with acute onset of dyspnea. He rapidly decompensated into PEA and was unable to be resuscitated. Autopsy showed intimal and medial thickening of his pulmonary arteries with marked luminal narrowing, consistent with pulmonary arterial hypertension (PAH).
There is one prior case report of two pediatric FHLH patients who died in the first year post-HSCT with PAH discovered on autopsy. Despite successful HSCT, there appears to be a pathologic link between FHLH and PAH, both early and late post-HSCT. PAH should be considered in any FHLH patient with respiratory symptoms and improved screening techniques are critically needed.