عنوان مقاله:

Indication Of Stem Cell Translantation In Nonmalignant Hematologic Disorders

محل انتشار:

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خلاصه مقاله:

Sickle cell disease(SCD)is the most common inherited hemoglobinopathy worldwide. The goal when performing HSCT is to replace the patient’s marrow with genetic functional cells before major organ dysfunction and complications it has been demonstrated that patients transplanted at a young age have a better 3-year OS and 3-year EFS, with lower incidence of aGvHD and cGvHDNonmalignant indications, however continue to grow, most importantly HCT for hemoglobinopathies by 36%, equally for thalassemia and sickle cell disease. In late 1990s, the Pesaro group has proposed a risk classification for pediatric patients undergo- ing MFD HSCT for TM . Limitations to this risk stratification include the interobserver variability regarding hepatomegaly and the lack of clear definition of adequate iron chelation. very-high- risk group was identified in Pesaro class 3 patients if liver size is > 5 cm below the costal margin and if the patient age is > 7 years. Transient elastography (FibroScan) and T2 MRI have been shown to be reliable noninvasive methods to predict liver fibrosis secondary to iron overload. effective and safe haploidentical transplant procedure for TM patients. The use of TCD graft was associated with high rate of infections and increased risk of graft failure due to allo-sensitization and hyperactive marrow . This was overcome by pretransplant over-transfusion and immunosuppressive therapy and post transplant infusion of transduced donor T-cells BU, CY, TT, and FLU as conditioning regimen and ATG HLA identical sibling HSCT is an estab-i lished treatment option for SCD. HSCT should be performed as early as possible, preferably at pre-school age, and BU, CY, and ATG should be used as conditioning regimen. Match family donor allo-HSCT is cur- rently considered the only curative standard therapeutic approach for thalas- semia major, which despite holding its own risks, could release the patient from lifelong treatments and possible iron accumulation complications. Despite encouraging results of gene therapy, its use in TM is currently limited to clinical trials. bone marrow failure syndrome Disease such as MDS, Myelofibrosis, hypocellular acute leukemia , inherited bone marrow failure and telomeropathies need to be excluded. Cytogenetic abnormalities can be found in up to 10% of true SAA.in case of transfusion requirement or if the criteria for SAA are met, treatment should begin with no delay. Early bone marrow HSCT after a conditioning regimen with CY, ATG, and ... GVHD prophylaxis combining CSA and MTX promotes excellent engraftment (95%) and OS (90% at 2 yea

کلمات کلیدی: