Objectives: Veno-occlusive disease (VOD) is a complication of hematopoietic stem cell transplantation (HSCT) known to develop before day 21 of HSCT, but VOD may develop later, termed late VOD. Mortality of VOD is high, but there is no proven definitive treatment. AT-III is an agent used in conventional VOD for its’ anti-inflammatory effect and the efficacy and safety of AT-III based therapy for late VOD was investigated. Methods: A retrospective analysis was done on 12 children who received AT-III for late VOD after HSCT at Seoul National University Children’s Hospital between June 2008 and June 2012. Three doses of 50 unit/kg/dose of AT-III were administered every 8 hours and further doses were administered every 24 hours. Prostaglandin E1 and low molecular weight heparin were used as prophylactic therapy and tissue-plasminogen activator (n=8) and defibrotide (n=2) were used as add-on therapy in some of the patients. Results: Patients underwent HSCT at median 11.5 years of age and late VOD was diagnosed at median 31 days (range, 21 to 54 days) after HSCT. AT-III at diagnosis of VOD was median 66%. Peak AT-III was median 198% after median 11 doses of AT-III, and final AT-III was median 135% after median 20 doses in median 18 days. VOD resolved in 10 patients (83%). Four patients died but only 1 patient (8%) died of VOD; others died of other causes (disease progression, septic shock, acute respiratory distress syndrome). Conclusion: AT-III based treatment may be an effective strategy for combating late VOD after HSCT.