

Figure S1. Flow of 249 patients in the HLH-94 treatment protocol

Within brackets are specifications of the fate of the patients at last follow-up. *: Out of 19 with a treatment change during the first 2 months, 18 received CSA (whereof 5 with familial disease known at onset). 3 CSA-treated patients died during the first 2 months of therapy. 1 patient changed to another treatment (familial, no HSCT donor found, deceased). 3 patients alive w/o HSCT (whereof 2 off all therapy >12 mo, considered secondary HLH and 1 with follow-up of <12 months). **: 4 out of these 32 patients had familial disease known at onset. §: 1 dead in surgical hemorrhage post-HSCT. §§: 1 stopped therapy due to varicella infection (dead). #: 1 patient with HSCT due to AML, not HLH. ##: 1 patient alive, without treatment for >12 months, considered secondary HLH. ♂: Follow-up of <12 months. ♂♂: 1 patient with follow-up of <12 months, 1 on a low, continuous dose of CSA due to reactivations. ♂♂♂: 1 patient with familial disease known at onset (HSCT, alive), 3 non-familial (0 HSCT, 3 dead).

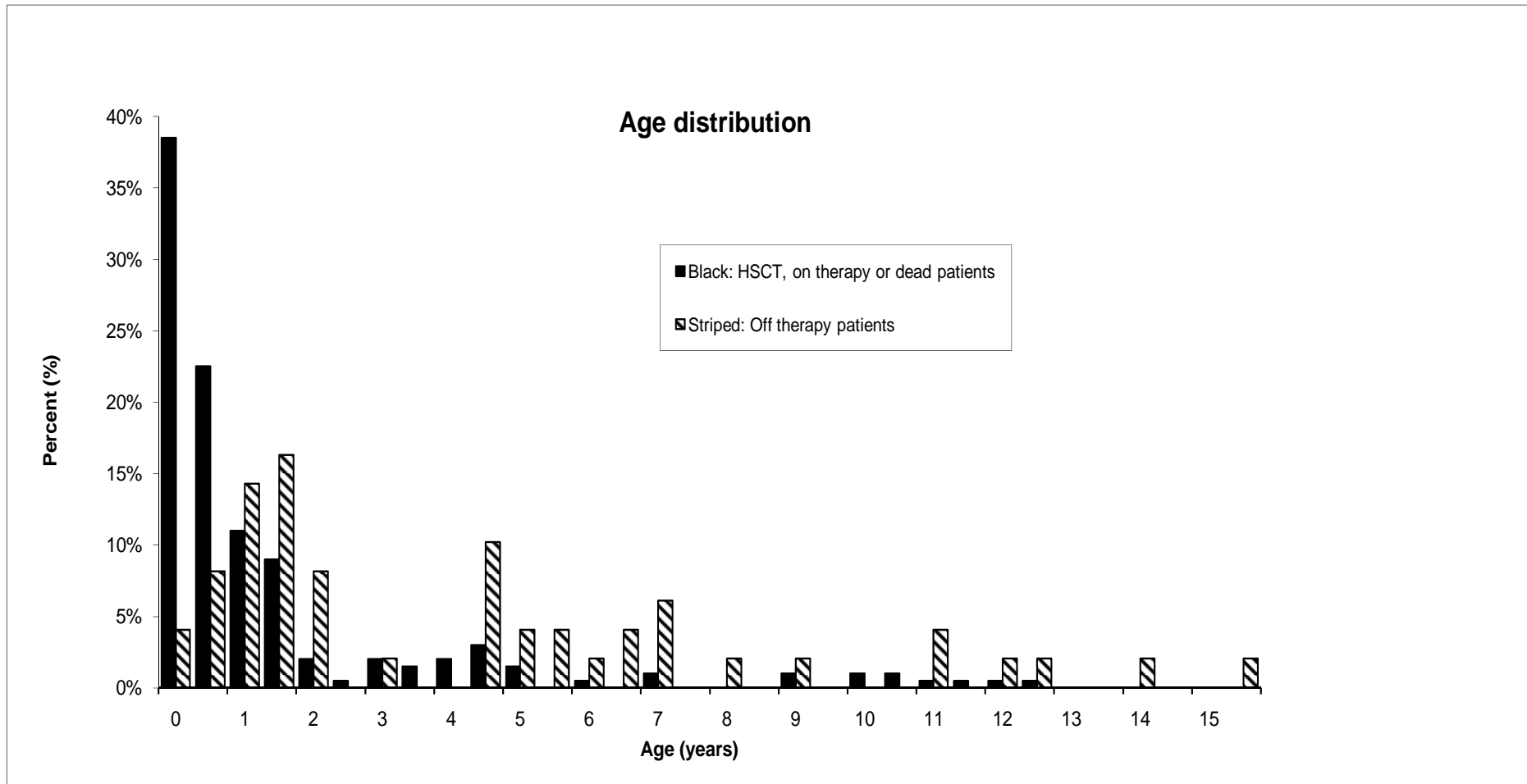


Figure S2. Comparison of age at start of HLH therapy between patients “off therapy” (= alive without HSCT and off all HLH therapy for at least a year with no signs of disease reactivation) and all the other patients ($p < 0.001$)