

Table E1: Outcome of cases with mixed chimerism

<u>ID</u>	<u>WAS Score</u>	<u>Transplant</u>	<u>Post-HSCT complications</u>	<u>Outcome/Follow-up</u>
P7	5	MUD,BM Alemtuzumab/Flu/Treo	Post-HSCT adenoviraemia Pneumatosis intestinalis Post-HSCT splenectomy (low PLT at 6m)	Off Ig. PIT=160 x10 ⁹ /L. Donor M: 0%. 83% T 105m
P8	2 Pre-HSCT splenectomy	MUD,PBSCs Alemtuzumab/Bu/Cyc	aGVHD grade II skin/gut	Off Ig. PIT=148x10 ⁹ /L. Donor Myeloid 7%80% T 105m
P12	2	MUD,BM Alemtuzumab/Flu/Mel	Grade IV skin aGvHD Adenoviral enteritis Recurrent AIHA/ITP/AIN Post-HSCT splenectomy	On Ig therapy (low B cell number) Bronchiectasis. PIT=116x10 ⁹ /L. Donor M: 0%. 74% T 138m
P16	5	MUD, PBSCs Alemtuzumab/Flu/Treo	On/off epistaxis	Off Ig. PIT=78 X10 ⁹ /L Donor myeloid 17%, 93% T 36m

P32	5	MUD, BM Alemtuzumab/Flu/Mel	Limited cGvHD	Off Ig. PIT=109x10 ⁹ /L. Donor M: 0%. T:74% 60m
		Pre-HSCT splenectomy		
P34	5	MUD, PBSCs Alemtuzumab/Flu/Treo	Recurrent vasculitis/autoinflammatory rash Patchy gut granulomas	On Ig therapy. PIT: 25x10 ⁹ /L. Donor M: 7%. 91% T 44m

Abbreviations: M: myeloid engraftment. PIT: platelets, Flu: Fludarabine, Treo: Treosulphan, Mel: Melphalan, FMF: familial Mediterranean fever, MUD: matched unrelated donor, BM: bone marrow, PBSCs: peripheral mononuclear stem cells, Ig: immunoglobulin, m: months, cGvHD: chronic GvHD.

Figure E1: Effect of donor T and myeloid engraftment and post-transplant autoimmunity

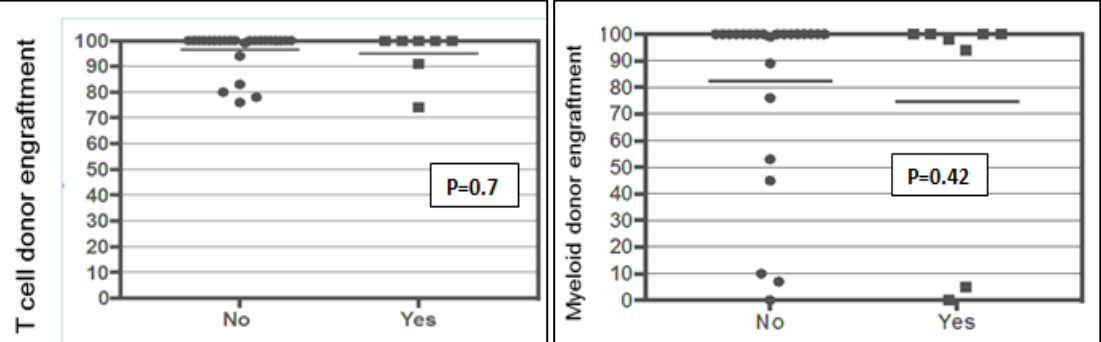
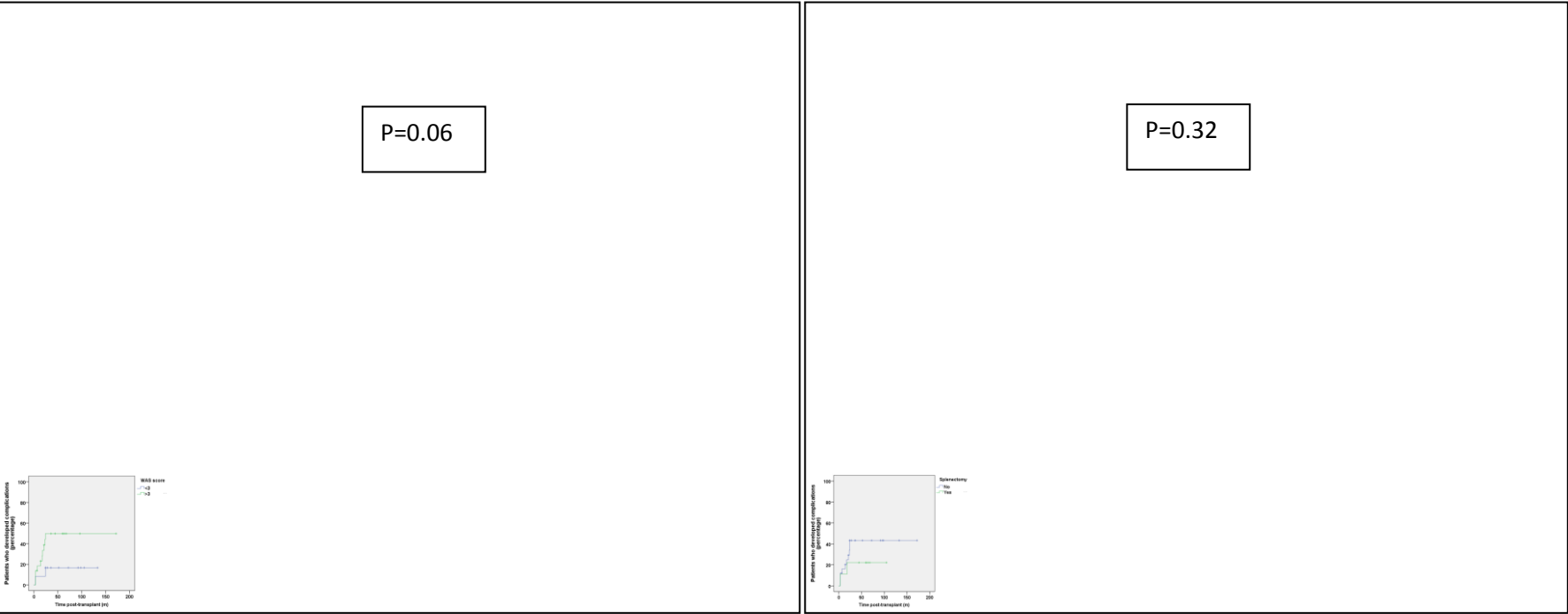


Figure E2: Probability of clinical and immunologic complications among 34 WAS patients



Complications included post-transplant autoimmunity, need for a second intervention, chronic GvHD, post-transplant splenectomy, reduced vision (post-CMV retinitis), bronchiectasis and secondary malignancy.