

Table 4 Published Transplanted MNGIE patients age more than 20 years^{12,13,14,15,16}

Age (years)	Donor/Stem cell source	Conditioning	Chimerism	Complications	Outcome
21	MUD/Cord	Flu/Cy/ATG	0%	Engraftment failure	Died from progression of MNGIE
21	MUD/BM	Bu/Flu	84%	AGVHD grade 1	Worsened GI symptoms Died on day +150 from intestinal perforation
22	MSD/BM	Bu/Flu/ATG	100%	Mild CGVHD	Died on day+246 from MNGIE related intestinal pseudoobstruction and septic shock
23	MRD/BM	Bu/Flu/ATG	Mixed chimerism	AGVHD grade 1	Died from MNGIE related perforation with infection
23	MRD/PBSC	Bu/Flu/MabC	73%		Alive with minimal improvement
23	MRD/BM	Bu/Flu	92%		Alive with some improvement
25	MUD/BM-1 st BMT MUD/PBSC	Bu/Flu/ATG Bu/Flu	1 st HSCT- 48% 2 nd HSCT- 100%	Secondary engraft. failure Moder. AGVHD	Alive
25	MRD/PBSC	Bu/Flu	100%	AGVHD grade 2 CGVHD	Alive on day +60 with worsened GI + neurology
25	MUD/BM	Bu/Flu	100%	AGVHD grade 2	Alive on day +283
26	MSD/BM	Bu/Flud	Engrafted day 16/19	Sepsis/Septic shock/MOF	Died on day +24 from infection
26	MUD/BM	Bu/Flu/ATG	100%	AGVHD grade 2	Alive
27	MRD/BM	Bu/Flu	Engrafted day14/13		Alive 14 mo post-transplant w/o improvement- progression of MNGIE
27	MUD/BM Unrelated Cord MM	Flu/Cy/ATG Bu/Flu/ATG	1 st HSCT - 0% 2 nd HSCT - 100%	1 st HSCT - Engraft failure Infection	Died after second HSCT from TRM, MOF and infection
29	Unrelated Cord MM	Cy/TBI/ATG	0%	AGVHD grade 2	Died on day +105 TRM (infection-reactivation of TB)
30	MSD/PBSC	Flu/Cy/ATG	100%		Alive with some clinical improvement
33	MUD/PBSC	Flu/Cy/ATG	0%	Engraftment failure	Died from progression of MNGIE
33	Unrelated double Cord	Flu/Cy/ATG	0% 1 st HSCT	1 st HSCT - Engraft failure	Needed 2 nd HSCT Died from AGVHD

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	MUD/PBSC		100% 2 nd HSCT	2 nd HSCT -AGVHD grade 4	
34	MMUD/BM	Bu/Flu	100%	GVHD GI stage 3-4 pancreatitis	Died on day +453 from worsening GI symptoms and sepsis due to perforation
34	MUD/BM	Bu/Cy	0%	VOD (postmortem liver cirrhosis)	Died on day+18 from VOD severe and MOF
34	MUD/BM	Bu/Flu/ATG	100%	AGVHD grade 3-4	Died from septic shock after intestinal perforation
40	Unrelated double Cord	Bu/Flu	100%	IPS	Died on day +21 TRM
41	MRD/PBSC	Bu/Flu/MabC	100%	Central line infection on day+1196	Died from infection

Abbreviations: MUD, matched unrelated donor; Flu, fludarabine; Cy, cyclophosphamide; ATG, anti-thymoglobulin; MNGIE, mitochondrial neurogastrointestinal encephalomyopathy; BM, bone marrow; Bu, busulfan; AGVHD, acute graft versus host disease; GI, gastrointestinal; MSD, matched sibling donor; CGVHD, chronic graft versus host disease; MRD, matched related donor; PBSC, peripheral blood stem cells; MOF, multi organ failure; TRM, transplant related mortality; MM, mismatched; MMUD, mismatched unrelated donor; TBI, total body irradiation; TB, tuberculosis; VOD, Hepatic veno-occlusive disease; IPS, idiopathic pneumonia syndrome; MabC, Alemtuzumab;