

Treatment	Dose	Patients	Number of treated patients/total of AIHA patients, line of treatment	Response rate Response/total of treated patients	First author, year
Splenectomy		Adults	2/15 as second line	1/2 CR	Sanz, 2014 ³¹
		Adults	2/19 as forth line	NR	Wang, 2015 ⁵
		Children	1/19 as third line	1/1 CR	Ahmed, 2015 ³⁰
		Adults	2/8 as second line	1/2 CR	Bhatt, 2016 ³²
		Children	2/12 as third line	1/2 CR	Kruizinga, 2018 ²¹
		Adults and children	7/60 (line of treatment not specified)	1/7 CR	Gonzalez-Vincent, 2018 ¹⁶
		Adults	1/10 as fifth line	1/1 CR	Scordo, 2019 ⁴²
Sirolimus		Adults	1/7	1/1 CR	Miller, 2020 ²⁹
	Not stated	Children	1 /15 as second line	1/1 PR	Faraci, 2014 ²⁷
	Not stated	Adults	1/19 as fifth line	NR	Wang, 2015 ⁵
	1 mg/m ² /day	1 child	1 as fourth line	1/1 CR, maintained at 2 years	Park, 2016 ⁴⁵
	Not stated	Children	1/12 as third line, 1/12 as forth line	2/2 CR	Kruizinga, 2018 ²¹
	Not stated	1 child, 1 adult	2	NR	Schuetz, 2018 ⁵⁰
	Not stated	Children	3/8 as third line (1 with RTX and steroids, 1 with steroids)	3/3 CR	Neely, 2019 ¹⁹
	Not stated	Children	2/12 as second line alone, 1 as second line with RTX and plasmapheresis, 1/12 as third line	2/2 CR as second line	Koo, 2020 ¹⁸
	Not stated	Children	3/19 as second/third line	3/3 CR with ongoing Sirolimus (+/- low dose steroids)	Lum, 2020 ²⁸
	Mycophenolate mofetil	Not stated	Adults	2/19 third and fourth line	1/2 CR
Not stated		Adults and children	11/60 (line of treatment not specified)	1/11 CR	Gonzalez-Vincent, 2018 ¹⁶
Not stated		Children	2/12 as second line	1/2 CR	Koo, 2020 ¹⁸
Bortezomib	1.3 mg/m ² /dose every 72 hour preceded by plasmapheresis	1 child	Fourth line	1/1 CR	Khandelwal, 2014 ⁶⁴
	1.3 mg/m ² IV at day 1, 4, 8, and 11 , 4 cycles	1 child	Third line	1/1 CR, persisting at 4 months after stopping Bortezomib	Mehta, 2014 ⁶⁵
	1.3 mg/m ² subcutaneously at day 1, 4, 8, and 11. Second cycle after 2 weeks	1 adult	Third line	1/1 CR	Cao, 2018 ⁶⁶

	1.3 mg/m ² /dose, once weekly for 1 to 4 weeks	Children	2/12 as second line, 2/12 as fourth line	2/4 CR	Kruizinga, 2018 ²¹
	Not stated	Adults and children	4/60 (line of treatment not specified)	1/4 CR	Gonzalez-Vincent, 2018 ¹⁶
	Not stated	1 adult	Second line	NR	Schuetz, 2018 ⁵⁰
	1.3 mg/m ² for 4 doses	1 child	Fourth line	NR	Even-Or, 2020 ⁶⁷
	Not stated	Children	2/19 as third line	NR: 1 died of infection, 1 needed 2 nd HSCT for refractory cytopenia	Lum, 2020 ²⁸
Cyclosporine	Not stated	Adults and children	2/20 as first line	2/2 CR	Daikeler, 2013 ²²
	Not stated	Adults	5/19 as third line (after steroids and RTX)	3/5 CR	Wang, 2015 ⁵
Daratumumab	16 mg/kg per week IV for 6 weeks	1 young adult, 2 children	>Fourth line	1/3 PR (patient then died), 2/3 CR (persisting 13 and 16 months after Daratumumab)	Schuetz, 2018 ⁵⁰
	16 mg/kg per week IV for 6 weeks	1 child	>fourth line	1/1 PR	Even-Or, 2020 ⁶⁷
	Not stated	Children	1/12 as third line, 1/12 as third line with Ofatumumab and Sirolimus	2/2 CR	Koo, 2020 ¹⁸
Abatacept	10 mg/kg every 2 weeks for 4 doses	Children	3 as third/fifth line	3/3 CR at 3, 5 and 9 months post Abatacept	Hess, 2018 ⁵¹
	10 mg/kg every 2 weeks for 4 doses	1 child	Third line	NR	Even-Or, 2020 ⁶⁷
IVIG alone	1 g/kg for 2 days	Adults	2/15 as first line	2/2 PR	Sanz, 2014 ³¹
	Not stated	Adults	1/19 as first line	1/1 CR	Wang, 2015 ⁵
	Not stated	Children	9/19 as first line	4/9 PR	Ahmed, 2015 ³⁰
	Not stated	Adults	2/10 as first line	1/2 CR	Scordo, 2019 ⁴²
	Not stated	Adults	1/7 as third line	NR	Miller, 2020 ²⁹
Plasma exchange		Adults	2/19 as second line	NR	Wang, 2015 ⁵
		Adults and children	7/60 (line of treatment not specified)	1/7 CR	Gonzalez-Vincent, 2018 ¹⁶
		Children	As second line: 1/12 alone, 1 with IVIG, 1/12 with Sirolimus and RTX	Response: alone NR, with IVIG CR, in combination PR	Koo, 2020 ¹⁸
Cyclophosphamide	Not stated	Children and adult	5/60 (line of treatment not specified)	2/5 CR	Gonzalez-Vincent, 2018 ¹⁶
	Not stated	children	1/8 as second line, 1/8 as third line	1/2 CR	Neely, 2019 ¹⁹
6-Mercaptopurine	Not stated	Children	3/15 as third line	3/3 CR	Chang, 2016 ³⁴
Azathioprine	Not stated	Adults	1/19 as third line	NR	Wang, 2015 ⁵
	Not stated	Children	1/8 as second line	NR	Neely, 2019 ¹⁹
Danazol	Not stated	Adults	1/19 as fourth line	1/1 PR	Wang, 2015 ⁵
	Not stated	Children	3/19 as third/fourth	2/3 CR	Ahmed, 2015 ³⁰

	Not stated	Children	1/8 as second line	NR	Neely, 2019 ¹⁹
Alemtuzumab	Not stated	Children and adult	1/60 (line of treatment not specified)	NR	Gonzalez-Vincent, 2018 ¹⁶
	15 mg/day IV three times per week	1 child	Fourth line	1/1 CR	Chao, 2008 ⁶⁸
Eculizumab	Not stated	Children and adults	2/60 (line of treatment not specified)	1/2 CR	Gonzalez-Vincent, 2018 ¹⁶
	Not stated	1 child	1/3	NR	Schuetz, 2018 ⁵⁰
Ofatumumab	Not stated	Children	1/12 as second line, 1/12 as third line with Daratumumab and Sirolimus	Second line: 1/1 CR, not known response to third line.	Koo, 2020 ¹⁸
Second HSCT		Children	1/12 as third line	NR	Kruizinga, 2018 ²¹
		Children	4/19	1/4 CR but died of toxicity, 3 had AIHA recurrence (1 had third HSCT, 1 responded to steroids, 1 died)	Ahmed, 2015 ³⁰
		Children	2/19 (HSCT for refractory cytopenia post AIHA)	2/2 CR	Lum, 2020 ²⁸

Table IV: Summary of relevant studies reporting different treatment modalities for post-HSCT AIHA beyond first line.

Abbreviations: 6-MP 6-mercaptopurine, AIHA autoimmune haemolytic anaemia, CR complete remission, CSA cyclosporine HSCT haematopoietic stem cell transplantation, IVIG Intra-venous Immunoglobulin, MMF mycophenolate mofetil, NR no response, PID paediatric immunodeficiencies, PR partial remission, RTX Rituximab