Supplementary table 1

1st line N=115	2nd line N=88	3rd line N=63	4th line N=33	5th line N=21	6th line N=19	7th line N=10	8th line N=10	9th line N=2
ITP N=41^	**- ITP* N=44**	**- ITP N=31**	**- ITP N=17*** **	**- ITP N=14**** **	**- ITP N=13**	**ITP N=4**	**- ITP N=5**	**- ITP N=1**
Steroids 26, 23 R	Steroids 18, 18 R	Steroids 13, 13 R	Steroids 5, 5 R	Steroids 3, 3 R	Steroids 8, 8 R	Steroids 4, 4 R	Steroids 2, 1 R	-
Steroids+IVIG 10, 8 R	Steroids+IVIG 8, 8 R	Steroids+IVIG 6, 5 R	Steroids+IVIG 2, 2 R	Steroids+IVIG 3, 3 R	Steroids+IVIG 3, 3 R	-	-	-
IST 3, 2 R	IST 5, 5 R	IST 3, 3 R	IST 3, 3 R	IST 1, 1 R	-	-	-	-
Danazol 1, 1 R	-	-		Danazol 1, 1 R	-	-	IST 2, 1 R	-
Rituximab 1, 1 R	Rituximab 5, 4 R	Rituximab 2, 2 R	Rituximab 3, 2 R	Rituximab 2, 2 R	-	-	-	-
Splenectomy 2, 2 R	Splenectomy 2, 2 OR	Splenectomy 2, 2 R	Splenectomy 2, 2 R	-	-	-	-	-
1 TPO-RA, 1 R	TPO-RA 8, 7 R	TPO-RA 5, 5 R	TPO-RA 3, 2 R	TPO-RA 5, 4 R	TPO-RA 2, 2 R	TPO-RA 1, 1 R	TPO-RA 2, 0 R	TPO-RA+ofatumumab, 1 R
- AIHA N=38^	**- AIHA* N=36**	**- AIHA N=26****	**- AIHA N=14**	**- AIHA N=6*****	**- AIHA N=6**	**- AIHA N=5**	**- AIHA N=5**	**- AIHA N=1**
Steroids 25, 21 R	Steroids 13, 13 R	Steroids 5, 5 R	Steroids 8, 8 R	Steroids 2, 2 R	-	Steroids 2, 2 R	Steroids 1, 1 R	-
Steroids+IVIG 3, 2 R	Steroids+IVIG 2, 1 R	Steroids+IVIG 3, 3 R	-	Steroids+IVIG 1, 1 R	-	-	-	-
Rituximab 3, 3 R	Rituximab 15, 14 R	Rituximab 10, 10 R	Rituximab 4, 4 R	Rituximab 2, 2 R	Rituximab 3, 3 R	Rituximab 1, 1 R	Rituximab 1, 0 R	-
IST 5, 5 R	IST 5, 5 R	IST 4, 3 R	IST 2, 2 R	IST 2, 2 R	IST 2, 2 R	IST 2, 1 R	IST 1, 1 R	IST 1, 1 R
Splenectomy 2, 2 R	Splenectomy 3, 3 R	Splenectomy 3, 3 R	-	Splenectomy 1, 1 R	-	-	-	-
Danazol 1, 1 R	-	Danazol 3, 2 R	-	-	-	-	Fostamatinib 1, 1 R	-
		Parsaclisib 1, 1 R	-	-	rEPO 1, 1 R	-	PEX 1, 1 R	-
- BOTH N=36^	**- BOTH N=8**	**- BOTH N=6**	**- BOTH N=2**	**-BOTH N=1**	**-BOTH N=0**	**- BOTH N=1**	**-BOTH N=0**	**-BOTH N=0**
Steroids 14, 11 R	Steroids 1, 1 R	-	-	-	-	-	-	-
Steroids+IVIG 10, 8 R	-	-	-	-	-	-	-	-
Rituximab 8, 7 R	Rituximab 6, 6 R	Rituximab 4, 4 R	Rituximab 2, 2 R	Rituximab 1, 1 R	-	-	-	-
IST 3, 3 R	IST 1, 1 R	-	-	-	-	-	-	-
TPO-RA 2, 2 R	-	TPO-RA 2, 2 R	-	-	-	TPO-RA + tocilizumab 1, 1 R	-	-
Danazol 1, 1 R	-	-	-	-	-	-	-	-
PEX 1, 1 R	-	-	-	-	-	-	-	-

Some patients received more than 1 therapy in the same line: ^1 ITP received splenectomy, rituximab and TPO-RA; 1 AIHA received IST and splenectomy, 1 concomitant AIHA/ITP received rituximab+IST, 2 TPO-RA+rituximab. *2 ITP treated with TPO-RA after splenectomy; 1 AIHA received IST, splenectomy and rituximab. ** 1 AIHA patient received danazol and rituximab; 2 AIHA patients received CTX and rituximab. ***1 ITP received rituximab and TPO-RA. **** 1 ITP received both IST and TPO-RA; 1 AIHA received rituximab, IST and splenectomy.

Supplementary table 2. Death rates (per 100 person-years) and hazard ratios of death according to age category and occurrence of intermediate time-dependent outcomes.

Time-dependent variable	No. subjects	No. deaths	Person-years	Death rate (per 100 person-years)	Crude HR	95% CI	Adjusted HR*	95% CI
Age (years)								
<65	81	7	771	0.9	1.00	Reference	1.00	Reference
65-79	27	9	145	6.2	8.30	2.75-25.0	5.47	1.72-17.4
80+	8	7	47	14.7	21.7	6.52-72.3	21.0	5.84-75.2
Relapse								
No	27	6	479	1.3	1.00	Reference	1.00	Reference
Yes	89	17	484	3.5	6.18	2.11-18.1	3.65	1.1.3-11.8
Infection								
No	78	11	853	1.3	1.00	Reference	1.00	Reference
Yes	38	12	110	10.9	10.3	4.31-24.7	6.76	2.63-17.4
Thrombosis								
No	91	16	903	1.8	1.00	Reference	1.00	Reference
Yes	25	7	60	11.6	6.85	2.69-17.4	6.13	1.99-18.9

CI, confidence interval; HR, hazard ratio
*Each variable adjusted for the other in a multivariable Cox regression model

Supplementary table 3. Multivariable analysis for overall mortality including only patients who died due to direct Evans syndrome complications. HR hazard risk. 95% CI confidence interval.

covariate	Hazard ratio	Standard Error	z	P	95% Conf. Interval	N. of deaths
Age >65 years	2.56	2.17	1.11	0.26	0.48-13.5	4
Age >80 years	1.17	1.70	0.11	0.91	0.06-20.4	1
Relapse	2.47	2.46	0.91	0.36	0.35-17.4	8
Infections	33.7	33.6	3.53	0.000	4.77-238.4	8
Thrombosis	15.5	14.5	2.94	0.003	2.49-96.9	4