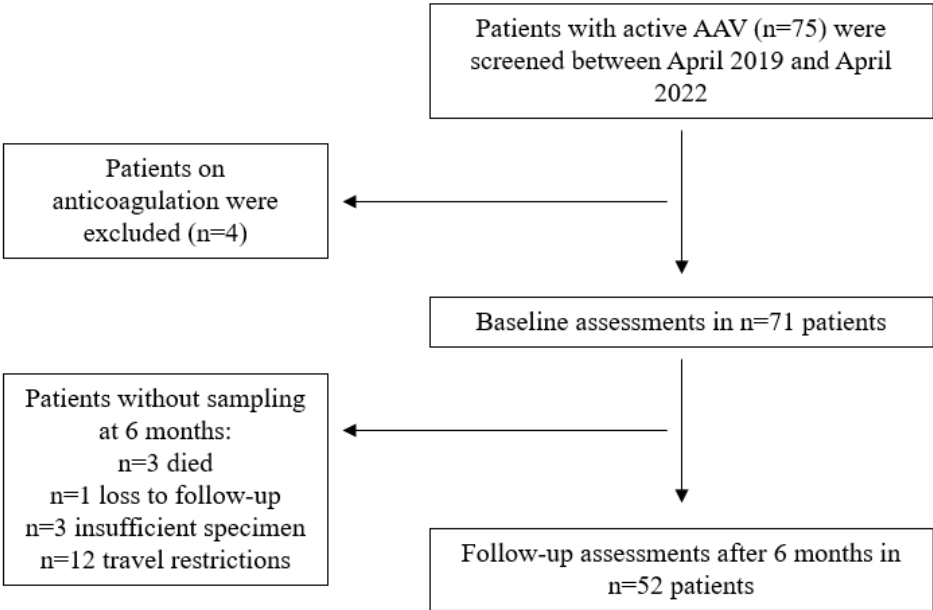


**Supplemental data**

**Figure 1.** Flowchart of the included patients and datapoint assessments.



Abbreviations: AAV, antineutrophil cytoplasmic antibody-associated vasculitis.

**Table 1.** Baseline characteristics of patients with and without thrombosis prior to inclusion.

	Normal range	Thrombosis n=4	No Thrombosis n=65	Overall <i>P</i>
Male, (%)		1 (25)	41 (63)	0.898
Age in years, (IQR)		63 (55-67)	64 (56-74)	0.130
BMI, (IQR)	18.5-25	30.5 (26.6-38.2)	25.7 (24.0-29.3)	0.053
Diagnosis, (%)				0.761
MPA		1 (25)	21 (32)	
GPA		3 (75)	44 (68)	
ANCA, (%)				0.697
MPO		1 (25)	28 (43)	
PR3		3 (75)	35 (54)	
Double positive		0 (0)	2 (3)	
De novo presentation, (%)		2 (50)	37 (57)	0.786
BVAS, (IQR)		14 (11-20)	14 (12-17)	0.949
Laboratory parameters				
Hemoglobin, (IQR)	8.2-11.0 <sup>†</sup> / 7.3-9.7 <sup>‡</sup> mmol/L	6.9 (5.6-8.8)	7.3 (6.2-8.0)	0.754
Platelet, (IQR)	130-350 x10 <sup>9</sup> /L	361 (282-435)	331 (263-389)	0.450
Leukocytes, (IQR)	3.5-11.0 x10 <sup>9</sup> /L	10.0 (6.8-15.0)	8.5 (6.8-12.1)	0.532
Serum creatinine, (IQR)	60-115 μmol/L	186 (72-425)	152 (95-229)	0.886
CRP, (IQR)	<10 mg/L	22 (13-68)	23 (7-45)	0.532
IgG, (IQR)	7.0-16.0 g/L	13.4 (8.8-14.3)	11.8 (9.1-13.2)	0.579
ANCA level, (IQR)		83 (47-113)	72 (30-144)	0.761
Coagulation parameters				
D-dimers, (IQR)	<5.0 <sup>#</sup> μg/mL	1.1 (0.3-1.8)	1.3 (0.6-2.7)	0.334
Fibrinogen, (IQR)	1.7-4.0 g/L	7.1 (5.0-9.6)	6.2 (5.2-8.5)	0.731
PT, (IQR)	10-12 sec	9.6 (8.5-17.4)	8.7 (8.3-9.2)	0.233
APTT, (IQR)	28-32 sec	26 (20-30)	27 (25-29)	0.397
FVIIa:AT, (IQR)	≤2641 pM	875 (792-909)	751 (481-916)	0.272
FXIIa:AT, (IQR)	≤118 pM	91 (29-2738)	59 (38-80)	0.835
FXIIa:C1Inh, (IQR)	≤2166 pM	4752 (3782-6566)	3889 (3562-4279)	0.221
FXIa:AT, (IQR)	≤43 pM	88 (62-412)	46 (25-93)	0.090
FXIa:α1AT, (IQR)	≤29 pM	62 (26-296)	26 (15-37)	0.115
FXIa:C1Inh, (IQR)	≤514 pM	269 (71-3654)	71 (71-209)	0.381
FIXa:AT, (IQR)	≤472 pM	191 (127-2509)	277 (197-364)	0.341
FXa:AT, (IQR)	≤279 pM	168 (145-464)	177 (164-206)	0.751
T:AT, (IQR)	≤4.7 μg/L	2.0 (1.7-32.3)	3.1 (1.8-6.3)	0.711

The Mann-Whitney U test was used for continuous variables and the chi-square test or Fisher's exact test was used for categorical variables, when appropriate. Normal hemoglobin ranges for <sup>†</sup>males and <sup>‡</sup>females. <sup>#</sup>D-dimers normal range for age 50 years or younger, with an incremental increase of 0.1 for each year above 50.

Abbreviations: SD, standard deviation. IQR, interquartile range. BMI, body mass index. MPA, microscopic polyangiitis. GPA, granulomatosis with polyangiitis. EGPA, eosinophilic granulomatosis with polyangiitis. ANCA, anti-neutrophil cytoplasmic antibody. MPO, myeloperoxidase. PR3, proteinase 3. BVAS, Birmingham vasculitis activity score. ENT, ear, nose and throat. RTX, rituximab. CYC, cyclophosphamide. CRP, C-reactive protein. IgG, immunoglobulin G. PT, Prothrombin time. aPTT, activated partial thromboplastin time. FVIIa, activated FVII. AT, antithrombin. FXIIa, activated factor XII. C1Inh, C1 esterase inhibitor. FXIa, activated factor XI. α1AT, α1-antitrypsin. FIXa, activated factor IX. FXa, activated factor X. T:AT, thrombin in complex with antithrombin.

**Table 2. Correlation matrix between clinical and laboratory findings and coagulation in patients with active AAV.**

	T:AT		D-dimers	
	Spearman's <i>r</i>	<i>P</i> value	Spearman's <i>r</i>	<i>P</i> value
Age in years	0.33	<b>0.006</b>	0.36	<b>0.003</b>
AAV type	0.05	0.688	- 0.05	0.683
ANCA type	0.12	0.337	- 0.01	0.938
De novo diagnosis	0.04	0.746	0.02	0.876
BVAS	0.12	0.325	0.35	<b>0.003</b>
Fibrinogen	- 0.09	0.475	0.08	0.523
PT	0.28	<b>0.018</b>	0.37	<b>0.002</b>
APTT	- 0.15	0.205	0.07	0.569
Hemoglobin	- 0.10	0.404	- 0.22	0.067
Platelet	0.15	0.213	0.13	0.294
Leukocytes	0.26	<b>0.033</b>	0.14	0.258
Serum creatinine	0.11	0.379	0.30	<b>0.014</b>
CRP	0.20	0.097	0.35	<b>0.003</b>
IgG	0.06	0.618	0.17	0.176
ANCA level	0.15	0.244	0.27	<b>0.028</b>
Proteinuria	0.29	<b>0.029</b>	0.40	<b>0.002</b>

Spearman rank correlation coefficients were used. Blue color indicates a positive correlation and red color indicates a negative correlation.

Abbreviation: AAV type, either microscopic polyangiitis or granulomatosis with polyangiitis. ANCA type, either myeloperoxidase or proteinase 3. BVAS, Birmingham vasculitis activity score. PT, Prothrombin time. aPTT, activated partial thromboplastin time. CRP, C-reactive protein. IgG, immunoglobulin G.

**Table 3.** Logistic regression was performed to ascertain the effects of routine clinical and coagulation parameters at baseline on the likelihood of developing a VTE in patients with AAV.

<b>Univariable</b>	<b>OR (95% CI)</b>	<b>P value</b>
Hemoglobin	1.735 (0.795-3.790)	0.167
Platelet	0.999 (0.990-1.007)	0.739
Leukocytes	1.028 (0.827-1.277)	0.802
Serum creatinine	0.995 (0.984-1.007)	0.443
CRP	1.000 (0.984-1.017)	0.981
IgG	0.916 (0.660-1.270)	0.598
ANCA level	0.994 (0.978-1.010)	0.451
D-dimers	1.105 (0.987-1.237)	0.083
Fibrinogen	0.658 (0.376-1.152)	0.143
PT	1.042 (0.628-1.729)	0.875
APTT	1.040 (0.906-1.194)	0.578
FVIIa:AT	0.999 (0.996-1.002)	0.502
FXIIa:AT	0.998 (0.986-1.009)	0.701
FXIIa:C1Inh	1.000 (0.999-1.001)	0.845
FXIa:AT	0.988 (0.959-1.017)	0.400
FXIa: $\alpha$ 1AT	0.999 (0.988-1.010)	0.795
FXIa:C1Inh	0.998 (0.994-1.003)	0.553
FIXa:AT	0.998 (0.993-1.004)	0.163
FXa:AT	0.965 (0.931-1.001)	0.054
T:AT	1.029 (0.949-1.115)	0.490

Abbreviations: 95% CI, 95% Confidence interval. CRP, C-reactive protein. IgG, immunoglobulin G. PT, Prothrombin time. aPTT, activated partial thromboplastin time. FVIIa, activated FVII. AT, antithrombin. FXIIa, activated factor XII. C1Inh, C1 esterase inhibitor. FXIa, activated factor XI.  $\alpha$ 1AT,  $\alpha$ 1-antitrypsin. FIXa, activated factor IX. FXa, activated factor X. T:AT, thrombin in complex with antithrombin.