

ONLINE SUPPLEMENTARY DATA

Supplementary Table 1. Patients' baseline characteristics at AAV diagnosis according to AAV subtypes and ANCA specificity

	MPA (n=27)	GPA (n=27)	EGPA (n=7)	P	MPO (n=39)	PR3 (n=13)	Negative (n=5)	Other (n=6)	p-value
Median age at AAV diagnosis, years, [IQR]	67 [59-77]	62 [49-73]	57 [43-76]	0.26	67 [55-76]	65 [60-70]	49 [45-57]	46 [27-73]	0.06
Sex (male), n (%)	6 (22)	8 (30)	2 (29)	0.8	11 (28)	3 (23)	1 (20)	1 (17)	1
Tabaco users, n (%) (N=56)	8 (30)	8 (36)	3 (43)	0.8	12 (32)	5 (56)	2 (40)	0 (0)	0.3
Prior autoimmune disease	8 (30)	4 (15)	0 (0)	0.2	10 (26)	2 (16)	0 (0)	(0)	0.5
Median duration between B and AAV diagnoses, year [IQR]	13 [3-54.5]	35 [16-48]	3 [3-3]	0.4	18.5 [3.75-56.75]	19.5 [14.5-26.5]	-	2 [2-2]	0.4
Median duration between AAV and B diagnoses, year [IQR]	0.5 [0-1]	5 [1.25-7.75]	1 [0.25-3.25]	0.04	1 [0-4]	4 [0-5]	1 [1-23]	6 [2.5-6]	0.8
AAV, n (%)									<0.001
GPA	-	-	-	-	11 (28)	13 (100)	1 (20)	2 (33.5)	
MPA	-	-	-	-	25 (64)	0 (0)	0 (0)	4 (66.5)	
EGPA	-	-	-	-	3 (8)	0 (0)	4 (80)	0 (0)	
ANCA specificity, n (%)									
MPO	25 (93)	11 (41)	3 (43)	<0.001	-	-	-	-	-
PR3	0 (0)	13 (48)	0 (0)	<0.001	-	-	-	-	-
Other*	4 (15)	2 (7)	0(0)	0.3	-	-	-	-	-
Negative,	0 (0)	1 (4)	4 (57)	<0.001	-	-	-	-	-
Symptoms and manifestations, n (%)									
Respiratory	20 (74)	20 (74)	5 (71)	1	28 (72)	10 (77)	4 (80)	3 (50)	1
ENT	6 (22)	22 (81)	4 (57)	<0.001	15 (38)	10 (77)	4 (80)	2 (33)	0.046
Neurologic	11 (41)	12 (44)	3 (43)	0.96	19 (49)	4 (31)	2 (40)	1 (17)	0.7
Mononeuritis multiplex	10 (37)	10 (37)	3 (43)	0.96	17 (44)	3 (23)	2 (40)	1 (17)	0.96
Renal	10 (37)	10 (37)	1 (14)	0.96	15 (38)	6 (46)	0 (0)	0 (0)	0.14
Proteinuria > 0.4 gm/24 hours	10 (37)	9 (33)	1 (14)	0.96	15 (38)	5 (38)	0 (0)	0 (0)	0.2

Creatinine >140 μ moles/liter	5 (18.5)	5 (18.5)	1 (14)	0.96	8 (20.5)	3 (23)	0 (0)	0 (0)	0.7
Myocarditis	0 (0)	1 (4)	3 (43)	0.004	2 (5)	0 (0)	2 (40)	0 (0)	0.06
Median BVAS [IQR]	11 [6-16]	16 [10-20]	14 [11-18]	0.2					
Median 1996 FFS [IQR]	0 [0-1]	0 [0-1]	1 [0-1]	0.4					
Treatments, n (%)									
Corticosteroids	27 (100)	26 (96)	7 (100)	1	38 (97)	13 (100)	5 (100)	4 (67)	1
Immunosuppressant	23 (84)	24 (89)	6 (86)	0.9	33 (85)	11 (85)	5 (100)	4 (67)	1
Cyclophosphamide	19 (70)	21 (78)	5 (71)	0.9	26 (67)	11 (85)	5 (100)	3 (50)	0.4
Rituximab	5 (18.5)	10 (37)	0 (0)	0.1	8 (21)	6 (46)	0 (0)	1 (17)	0.1
Azithromycin	8 (30)	2 (7)	2 (29)	0.09	10 (26)	1 (8)	1 (20)	0 (0)	0.4

*: 2 patients are both anti-MPO (myeloperoxidase) and anti-BPI (bactericidal/permeability-increasing protein) positive

AAV, ANCA associated vasculitis; ANCA, antineutrophil cytoplasmic antibodies; B, bronchiectasis; BVAS, Birmingham Vasculitis Activity Score; EGPA, eosinophilic granulomatosis with polyangiitis; ENT, ear nose and throat; FFS, five factor score; GPA, granulomatosis with polyangiitis; IQR, interquartile range; MPA, microscopic polyangiitis; MPO, myeloperoxidase; PR3, proteinase 3.

Supplementary Table 2. Respiratory findings at the diagnosis of bronchiectasis according to AAV subtypes and ANCA specificity

	MPA (n=27)	GPA (n=27)	EGPA (n=7)	p	MPO (n=39)	PR3 (n=13)	Negative (n=5)	Other (n=6)	p- value
Median age at B diagnosis, years, [IQR]	59 [43-71.5]	56 [41.5-72]	60 [51.5-73]	0.7	56 [32-73]	60 [50-70]	60 [58-66]	51 [41.25-63.25]	0.7
Respiratory manifestations, n (%)	20 (74)	20 (74)	5 (71)	1	28 (72)	10 (77)	4 (80)	3 (50)	1
Dyspnea	12 (44)	12 (44)	5 (71)	0.5	19 (49)	6 (46)	3 (60)	1 (17)	0.4
Cough	16 (59)	12 (44)	4 (57)	0.6	20 (51)	6 (46)	3 (60)	3 (50)	0.7
Hemoptysis	8 (30)	7 (26)	1 (14)	0.8	10 (26)	4 (31)	1 (20)	1 (17)	0.9
FACED score (0-7), median [IQR], (N=32)	3 [2-4]	1.5 [1-4.5]	2 [1-4]	0.7	3 [2-5]	1 [0-1]	1 [1-3]	3 [3-4]	0.2
Bhalla's score (0-25), median [IQR] (N=53)	10 [8-12]	9 [6-12]	7 [5-8]	0.1	10 [7-12]	9 [6.25-9.75]	8 [4-8]	10 [5.5-13.5]	0.4
Other radiologic findings, n (%)									
Nodules	7 (26)	9 (33)	3(43)	0.6	12 (31)	5 (38)	1 (20)	1 (17)	0.6
Nodule excavation	0 (0)	2 (7)	0 (0)	0.4	0 (0)	2 (16)	0 (0)	0 (0)	0.07
Intra alveolar hemorrhage	2 (7)	0 (0)	0 (0)	0.6	2 (5)	0 (0)	0 (0)	0 (0)	1
Interstitial lung disease	1 (3.5)	0 (0)	0 (0)	1	1(2.5)	0 (0)	0 (0)	0 (0)	1
Respiratory function									
FEV1, ml median [IQR] (N=34)	1430 [1097-2000]	1810 [1170-2080]	1090 [1070-2590]	1	1500 [1090-2020]	1780 [1315-1817]	1200 [1060-1895]	1635 [1032-2185]	1
FEV1 %, median [IQR] (N=34)	79 [59-88]	68 [61-75]	87 [54-104.75]	0.3	83 [59-93]	67 [63.5-70.75]	80 [64.25-102.75]	67.5 [50-81]	0.5
FEV1/FVC %, median [IQR] (N=40)	73 [69-81]	66 [64-73]	67 [51.5-71.25]	0.1	73 [66.5-80]	64 [62-66]	68 [56.5-80.5]	68.5 [64.5-69]	0.2
TPC ml, median [IQR] (N=31)	4970 [4210-5340]	6425 [4450-7225]	5980 [5395-6700]	0.05	5330 [4720-6340]	5390 [3857-6787]	5525 [5037-6012]	5200 [4810-6250]	1
TPC %, median [IQR] (N=31)	91 [77-97]	97 [94-101]	119 [109-127.5]	0.01	97 [89-110]	93 [90.25-95.75]	102 [101.5-102.5]	100 [97-104.5]	0.6

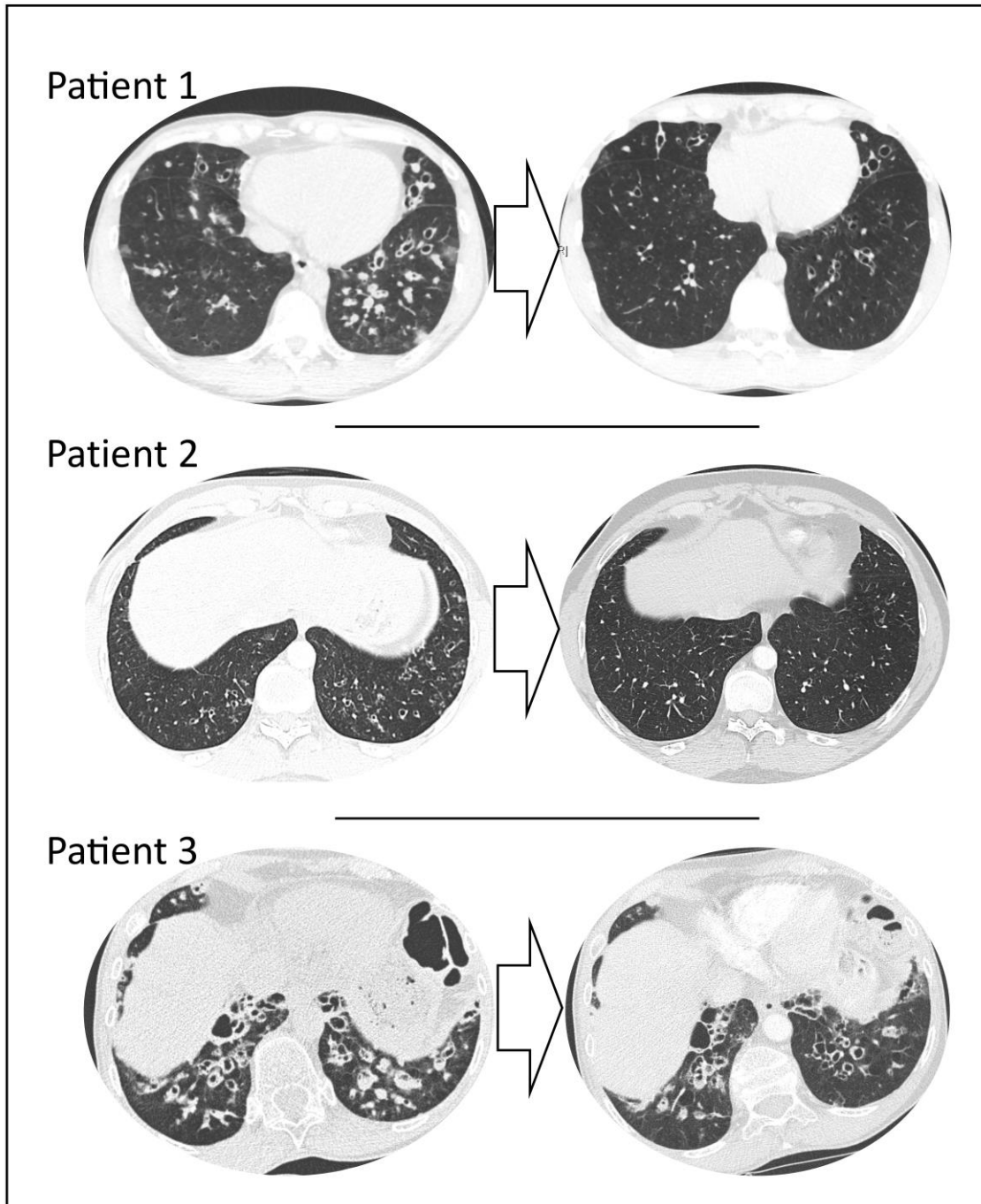
AAV, ANCA associated vasculitis; EGPA, Eosinophilic Granulomatosis with Polyangiitis; FACED, FEV1, Age, Chronic colonization, Extension and Dyspnea; FVC, forced vital capacity; FEV1, forced expiratory volume; GPA, granulomatosis with polyangiitis; IQR, interquartile range; MPA, microscopic polyangiitis; MPO, myeloperoxidase; PR3, proteinase 3; TPC, total pulmonary capacity.

Supplementary Table 3. Full detail of the Bhalla score at the diagnosis of bronchiectasis

First diagnosis	All (n=61)	B -AAV (n=25)	AAV-B (n=36)	p-value
Severity of bronchiectasis (0-3), n (%)				0.3
0-1 (absent to mild)	24 (45)	8 (35)	16 (53)	
2-3 (moderate to severe)	29 (55)	15 (65)	14 (47)	
Peribronchial thickening (0-3), n (%)				0.8
0-1 (absent to mild)	39 (74)	16 (70)	23 (77)	
2-3 (moderate to severe)	14 (26)	7 (30)	7 (23)	
Bronchiectasis extent (0-3), n (%)				0.3
0-1 (<6 BP segments)	26 (49)	9 (39)	17 (57)	
2-3 (≥6 BP segments)	27 (51)	14 (61)	13 (43)	
Mucous extension (0-3), n (%)				0.6
0-1 (<6 BP segments)	48 (91)	20 (87)	28 (93)	
2-3 (≥6 BP segments)	5 (9)	3 (13)	2 (7)	
Sacculation and abscesses (0-3), n (%)				0.07
0-1 (<6 BP segments)	50 (94)	20 (87)	30 (100)	
2-3 (≥6 BP segments)	3 (6)	3 (13)	0 (0)	
Generations of bronchial divisions involved (0-3), n (%)				1
0-1 (up to 4 th generation)	7 (13)	3 (13)	4 (13)	
2-3 (more than 4 th generation)	46 (87)	20 (87)	26 (87)	
Number of Bullae (0-3), n (%)				0.2
0 (absent)	48 (91)	19 (83)	29 (97)	
1-3 (unilateral, bilateral and >4)	5 (5)	4 (17)	1 (3)	
Emphysema (0-2), n (%)				0.7
0 (absent)	35 (66)	14 (61)	21 (70)	
1 (1-5 BP segments)	14 (26)	7 (30)	7 (23)	
2 (>5 BP segments)	4 (8)	2 (9)	2 (7)	
Consolidation (0-2), n (%)				0.8
0 (absent)	33 (62)	13 (57)	20 (67)	
1 (subsegmental)	12 (23)	6 (26)	6 (20)	
2 (segmental/lobar)	8 (15)	4 (17)	4 (13)	

AAV, ANCA associated vasculitis; AAV-B, bronchiectasis post vasculitis; ANCA, antineutrophil cytoplasmic antibodies; B-AAV, bronchiectasis before vasculitis; BP, bronchopulmonary

Supplementary Figure 1. Chest CT-scans of 3 patients with bronchiectasis occurring after onset of ANCA-associated vasculitis before (left panel) and after (right panel) immunosuppressive therapy.



Supplementary Table 4. Patients’ outcome according to AAV subtypes and ANCA specificity

	MPA (n=27)	GPA (n=27)	EGP A (n=7)	P	MPO (n=39)	PR3 (n=13)	Negative (n=5)	Other (n=6)	p- value
AAV Follow-up (years), median [IQR]	5 [4-10]	10 [7-16]	3 [2-10]	0.00 7	6 [4-10]	10 [6-12]	3 [3-27]	18.5 [14-23]	0.08
Relapse of AAV, n (%)	10 (37)	13 (48)	3 (43)	0.6	15 (38)	6 (46)	2 (40)	3 (50)	0.6
Median number of AAV relapses [IQR]	0 [0-1]	0 [0-2]	0 [0-1]	0.4	0 [0-1]	0 [0-2]	0 [1]	1 [0.75-1.75]	0.4
Death, n (%)	8 (30)	1 (4)	0 (0)	0.02	7 (22)	1 (8)	0 (0)	1 (17)	0.5
Overall survival during AAV follow-up									
5 years, %	86	100	100		95	100	100	100	
10 years, %	56	92	100		81	75	-	-	
Median absolute variation of Ballah’s score, point [IQR]	1 [0-4]	2 [0-6]	1 [0-11]	0.83	3 [0-2.5]	0.5 [-1 – 2.25]	5.5 [2.75-8.25]	0 [-0.5 - 0.5]	0.4
Variation of Bhalla’s score, n (%) (N=37)				1					0.9
Degradation	11 (58)	9 (60)	2 (67)		16 (64)	4 (50)	1 (50)	1 (50)	
Stability	5 (26)	3 (20)	1 (33)		7 (28)	1 (12.5)	1 (50)	0 (0)	
Improvement	3 (16)	3 (20)	0 (0)		2 (8)	3 (37.5)	0 (0)	1 (50)	
Median annual infection rate [IQR], (N=16)	0.1 [0-0.7]	0.2 [0-0.4]	0 [0-0.2]	0.5	0 [0-0.4]	0.2 [0-0.3]	0.2 [0-0.55]	0.45 [0.2-0.8]	0.7
<i>Pseudomonas aeruginosa</i>’s colonization, n (%)	9 (33)	8 (29.5)	1 (14)	0.8	9 (23)	5 (38)	2 (40)	2 (33)	0.4

AAV, ANCA-associated vasculitis; ANCA, antineutrophil cytoplasmic antibodies; EGPA, Eosinophilic Granulomatosis with Polyangiitis; GPA, granulomatosis with polyangiitis; IQR, interquartile range; MPA, microscopic polyangiitis; MPO, myeloperoxidase; PR3, proteinase 3.