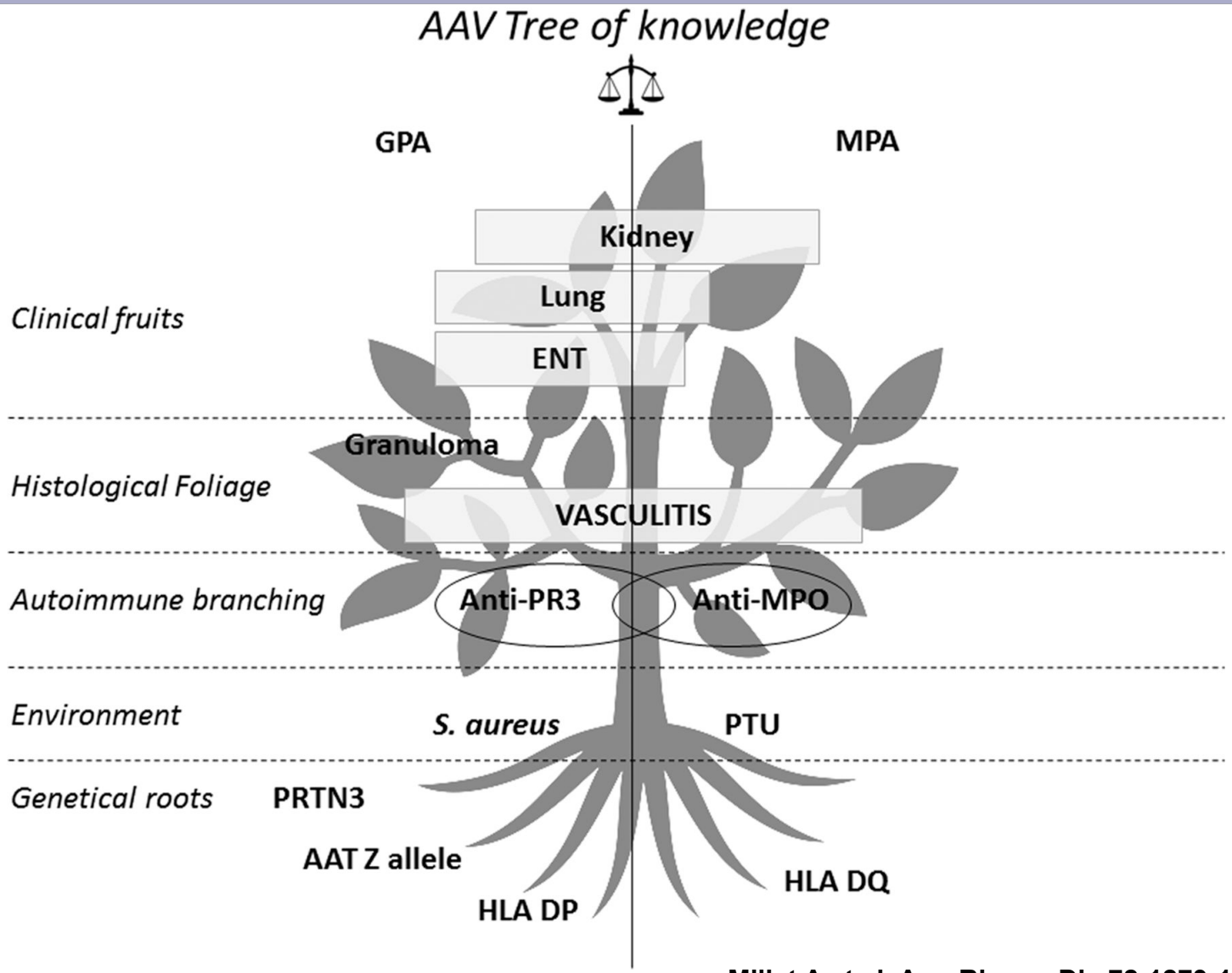
ANCA-assocated Vasculitis (AAV)



Millet A et al. Ann Rheum Dis 72:1273-1279 (2013)

Compared features of granulomatosis with polyangiitis (GPA), and microscopic polyangiitis (MPA). We compare GPA and MPA in all the different aspects from genetics to pathophysiological characteristics and clinical manifestations, in order to illustrate the major differences between these two conditions. ANCA, antineutrophil cytoplasmic antibodies; AAT, α1-antitrypsin; AAV, ANCA-associated vasculitis; ENT. ear nose throat; HLA, human leukocyte antigen; MPO, myeloperoxidase; PR3, proteinase 3; PRTN3, gene coding for PR3; PTU, propylthiouracil.

ANCA-associated vasculitis (AAV)

Epidemiological, clinical and biological characteristics of patients with granulomatosis with polyangiitis (GPA), microscopic polyangiitis (MPA) and eosinophilic GPA (EGPA).11–20

	GPA	MPA	EGPA	
Gender	Male>female	Male>female	Male=female	
Peak disease, age (years)	64–74	64–74	40–60	
Geographical distribution	Equator to pole (in Europe)	Pole to equator (in Europe)	NA	
Pattern of appearance	Cyclical	Random	NA	
Environmental factors pharmacological induction	Silica exposure *Staphylococcus aureus*	Silica exposure Propylthiouracil	Silica exposure Leukotriene receptors antagonists	
			ANCA+	ANCA−
Organ involvement (%)				
Skin	30–60	40–70	53–67	51–62
Kidneys	50–80	90–100	31–51	4–16
Lungs	60–80	20–60	34–56	60–76
Ear, nose, throat	80–90	20–30	60–77	53–78
Joints	50–80	30–70	30–40	30–40
Peripheral nervous system	10–50	20–30	71–84	42–65
Eye	30–60	10–30	<10	<10
Bowel	<10	10–30	0–42	14–26
Heart involvement	5–15	10–20	0–12	22–49
Histopathological findings	Necrotising vasculitis and granulomas	Necrotising vasculitis	Necrotising vasculitis Eosinophilic granulomas	
PR3-ANCA	s-GPA 70–95% l-GPA 40–50%	10–20%	0–3.2%	
MPO-ANCA	0–10%	30–80%	32–92%	
Genetic association GWAS (2012)	AAT deficiency HLA DP PRTN3	HLA DQ	HLA-DRB4	
Major associations from previous studies	AAT Z allele PRTN3§ HLA DPB1*0401 PTPN22‡CTLA4‡	PTPN22‡ CTLA4‡	IL-10 promotor (ANCA-subset)	

§Single nucleotide polymorphisms different from Genome Wide Association Study (GWAS).

‡Not confirmed by the GWAS.

ANCA, antineutrophil cytoplasmic antibody; GPA, granulomatosis with polyangiitis; s-GPA, systemic GPA; l-GPA, localised GPA; EGPA, eosinophilic granulomatosis with polyangiitis; HLA, human leukocyte antigen; MPA, microscopic polyangiitis; NA, non applicable.

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