

**Supplementary Table 1.** Application of the 2007 EMA Algorithm for AAV and cPAN Based on the 2012 CHCC Definitions to 18 BD Patients with ANCA

Patients number	ANCA positivity	ACR for EGPA	ACR for GPA	Histology compatible with CHCC MPA and GPA surrogate markers	No histology GPA surrogate markers and PR3-or MPO-ANCA positivity	Clinical and histologic evidence of small-vessel vasculitis No GPA surrogate markers	No histology No GPA surrogate markers PR3- or MPO-ANCA positivity and renal vasculitis	Histology compatible with CHCC cPAN or typical angiographic features of cPAN	Not compatible with AAV or cPAN
	Reclassified as	EGPA	GPA	GPA	GPA	GPA	MPA	cPAN	Unclassified*
1	MPO(P)-ANCA	No	No	No	No	No	No	No	Yes
2	MPO(P)-ANCA	No	No	No	No	No	No	No	Yes
3 <sup>†</sup>	MPO(P)-ANCA	No	No	No	No	No	No	No	Yes
4	MPO(P)-ANCA	No	No	No	No	No	No	No	Yes
5	MPO(P)-ANCA	No	No	No	No	No	No	No	Yes
6 <sup>†</sup>	PR3(C)-ANCA	No	No	No	No	No	No	No	Yes
7 <sup>†</sup>	MPO(P)-ANCA	No	No	No	No	No	No	No	Yes
8	PR3(C)-ANCA	No	No	No	No	No	No	No	Yes
9	PR3(C)-ANCA	No	No	No	No	No	No	No	Yes
10 <sup>†</sup>	MPO(P)-ANCA	No	No	No	No	No	No	No	Yes
11	MPO(P)-ANCA	No	No	No	No	No	No	No	Yes
12	MPO(P)-ANCA	No	No	No	No	No	No	No	Yes
13	MPO(P)-ANCA	No	No	No	No	No	No	No	Yes
14 <sup>†</sup>	MPO(P)-ANCA	No	No	No	No	No	No	No	Yes
15	MPO(P)-ANCA	No	No	No	No	No	No	No	Yes
16	PR3(C)-ANCA	No	No	No	No	No	No	No	Yes
17	PR3(C)-ANCA	No	No	No	No	No	No	No	Yes
18	PR3(C)-ANCA	No	No	No	No	No	No	No	Yes

EMA, European Medicine Agency; AAV, antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis; cPAN, classic polyarteritis nodosa; CHCC, Chapel Hill Consensus Conference; ANCA, antineutrophil cytoplasmic antibody; ACR, American College of Rheumatology; EGPA, eosinophilic granulomatosis with polyangiitis; GPA, granulomatosis with polyangiitis; MPA, microscopic polyangiitis; PR3, proteinase 3; MPO, myeloperoxidase.

\*Since eighteen patients met the International study group (1990) and the International criteria for Behçet's disease (2014) criteria for Behçet's disease, all patients should be classified as Behçet's disease rather than unclassified vasculitis, <sup>†</sup>These five patients did not satisfy the ISG (1990) criteria, but only satisfy the ICB (2014) criteria.