Supplementary material for Sada K.-E. et al. A nationwide survey on the epidemiology and clinical features of eosinophilic granulomatosis with polyangiitis (Churg-Strauss) in Japan. Mod Rheumatol. 2014;24(4):640–4.

Supplementary Appendix. The 2nd survey questionnaire for patients with eosinophilic granulomatosis with polyangiitis.

I . Information of investigator	
1. Institution:	
2. Location:	3. Departments:
4. Name of investigator:	5. Screening date(YYYY/MM/DD) :
6. Patient ID:	
7. Date of onset of first symptom (YYYY/MM/DD):	8. Date of first visit of your institution (YYYY/MM/DD):
9. Date of definite diagnosis(YYYY/MM/DD):	10. Institution of definite diagnosis (0: Your institution, 1: Other institution, 9: Unknown)
II . Patients backgrounds	
1. Date of birth (YYYY/MM/DD):	2. Residential prefecture
3. Sex	1:Male, 2:Female
4. Allergic complications at definite diagnosis (Exclude seasonal or transient disease) 0:No, 1:Yes, 9:Unknown	
4-1. If yes, specify the diseases	1:Bronchial asthma (Date of onset YYYY/MM/DD:)
	2:Parenial allergic rhinitis (Date of onset YYYY/MM/DD:)
	3:Other disease (Specify the disease) (Date of onset YYYY/MM/DD:)
5. History of corticosteroids use for allergic disease	0:No, 1:Internal use, 2:Inhalant, 9:Unknown
6. Drug allergy	0:No, 1:Yes, 9:Unknown
6-1 If yes, specify the drug	
7. History of use of leukotriene receptor antagonist	0:No, 1:Yes, 9:Unknown
7-1 If yes, specify the drug	1:Pranlukast, 2:Zafirlukast, 3:Montelukast, 4:Others (Specify the drug)
8. Familial history of vasculitis	0:No, 1:Yes, 9:Unknown
8-1 If yes, specify the disease	1:Eosinophilic granulomatosis with polyangiitis, 2:Microscopic polyangiitis, 3:Granulomatosis with polyangiitis, 4:Other disease (Specify the disease)
8-2 Relationship	1:Father, 2:Mother, 3:Child, 4:Sibling, 5:Grandparent, 6:Grandchild
III. Histological examination	
1. Biopsy	0:Not performed, 1:Performed, 9:Unknown
1-1 If performed, specify the site	1:Skin, 2:Paranasal sinus, 3:Lung, 4:Kidney, 5:Peripheral nerve 6:Others (Specify the site)
1-2 Findings	1:Infiltration of eosinophil to extravascular tissue, 2:Necrotizing vasculitis, 3:Extravascular granulomas, 4:Others (Specify the findings)
1-3 Clinical course until onset	1:Characteristic (Symptoms due to vasculitis follow allergic disease and eosinophilia), 2:Not characteristic

IV. Clinical findings (「/」 means 「OR」)	A. At definite diagnosis	B. At this screening
1. General		
Fever (> 38.0 °C)	0: Absent, 1: Present, 9: Unknown	0: Absent, 1: Present within 4 weeks, 2: Persistent over 4 weeks, 9: Unknown
Weight loss (>2kg)	0: Absent, 1: Present, 9: Unknown	0: Absent, 1: Present within 4 weeks, 2: Persistent over 4 weeks, 9: Unknown
Myalgia	0: Absent, 1: Present, 9: Unknown	0: Absent, 1: Present within 4 weeks, 2: Persistent over 4 weeks, 9: Unknown
Arthralgia	0: Absent, 1: Present, 9: Unknown	0: Absent, 1: Present within 4 weeks, 2: Persistent over 4 weeks, 9: Unknown
2. Cutaneous		
Infarct	0: Absent, 1: Present, 9: Unknown	0: Absent, 1: Present within 4 weeks, 2: Persistent over 4 weeks, 9: Unknown
Purpura	0: Absent, 1: Present, 9: Unknown	0: Absent, 1: Present within 4 weeks, 2: Persistent over 4 weeks, 9: Unknown
Ulcer	0: Absent, 1: Present, 9: Unknown	0: Absent, 1: Present within 4 weeks, 2: Persistent over 4 weeks, 9: Unknown
Gangrene	0: Absent, 1: Present, 9: Unknown	0: Absent, 1: Present within 4 weeks, 2: Persistent over 4 weeks, 9: Unknown
Other skin vasculitis	0: Absent, 1: Present, 9: Unknown	0: Absent, 1: Present within 4 weeks, 2: Persistent over 4 weeks, 9: Unknown
3. Mucous membranes/Eyes		
Mouth ulcers/ Mouth granulomas	0: Absent, 1: Present, 9: Unknown	0: Absent, 1: Present within 4 weeks, 2: Persistent over 4 weeks, 9: Unknown
Genital ulcers	0: Absent, 1: Present, 9: Unknown	0: Absent, 1: Present within 4 weeks, 2: Persistent over 4 weeks, 9: Unknown
Sialadenitis	0: Absent, 1: Present, 9: Unknown	0: Absent, 1: Present within 4 weeks, 2: Persistent over 4 weeks, 9: Unknown
Dacryoadenitis	0: Absent, 1: Present, 9: Unknown	0: Absent, 1: Present within 4 weeks, 2: Persistent over 4 weeks, 9: Unknown
Proptosis	0: Absent, 1: Present, 9: Unknown	0: Absent, 1: Present within 4 weeks, 2: Persistent over 4 weeks, 9: Unknown
Episcleritis / Scleritis	0: Absent, 1: Present, 9: Unknown	0: Absent, 1: Present within 4 weeks, 2: Persistent over 4 weeks, 9: Unknown
Conjunctivitis / Blepharitis / Keratitis	0: Absent, 1: Present, 9: Unknown	0: Absent, 1: Present within 4 weeks, 2: Persistent over 4 weeks, 9: Unknown
Blurred vision	0: Absent, 1: Present, 9: Unknown	0: Absent, 1: Present within 4 weeks, 2: Persistent over 4 weeks, 9: Unknown
Sudden visual loss	0: Absent, 1: Present, 9: Unknown	0: Absent, 1: Present within 4 weeks, 2: Persistent over 4 weeks, 9: Unknown
Uveitis	0: Absent, 1: Present, 9: Unknown	0: Absent, 1: Present within 4 weeks, 2: Persistent over 4 weeks, 9: Unknown
Retinal change (vasculitis, thrombosis, exudate, haemorrhage)	0: Absent, 1: Present, 9: Unknown	0: Absent, 1: Present within 4 weeks, 2: Persistent over 4 weeks, 9: Unknown
4. Ear, Nose, and Throat		
Bloody nasal discharge / crusts / ulcers / granulomata	0: Absent, 1: Present, 9: Unknown	0: Absent, 1: Present within 4 weeks, 2: Persistent over 4 weeks, 9: Unknown
Paranasal sinus involvement	0: Absent, 1: Present, 9: Unknown	0: Absent, 1: Present within 4 weeks, 2: Persistent over 4 weeks, 9: Unknown
Subglottic stenosis	0: Absent, 1: Present, 9: Unknown	0: Absent, 1: Present within 4 weeks, 2: Persistent over 4 weeks, 9: Unknown
Conductive hearing loss	0: Absent, 1: Present, 9: Unknown	0: Absent, 1: Present within 4 weeks, 2: Persistent over 4 weeks, 9: Unknown
Sensorineural hearing loss	0: Absent, 1: Present, 9: Unknown	0: Absent, 1: Present within 4 weeks, 2: Persistent over 4 weeks, 9: Unknown

5. Chest		
Wheeze	0:Absent,1:Present,9:Unknown	0:Absent,1:Present within 4 weeks, 2:Persistent over 4 weeks, 9: Unknown
Nodules / Cavities	0:Absent,1:Present,9:Unknown	0:Absent,1:Present within 4 weeks, 2:Persistent over 4 weeks, 9: Unknown
Pleural effusion / Pleurisy	0:Absent,1:Present,9:Unknown	0:Absent,1:Present within 4 weeks, 2:Persistent over 4 weeks, 9: Unknown
Infiltrate	0:Absent,1:Present,9:Unknown	0:Absent,1:Present within 4 weeks, 2:Persistent over 4 weeks, 9: Unknown
Ground glass opacity / Interstitial pneumonitis	0:Absent,1:Present,9:Unknown	0:Absent,1:Present within 4 weeks, 2:Persistent over 4 weeks, 9: Unknown
Endobronchial involvement	0:Absent,1:Present,9:Unknown	0:Absent,1:Present within 4 weeks, 2:Persistent over 4 weeks, 9: Unknown
Massive haemoptysis / alveolar haemorrhage	0:Absent,1:Present,9:Unknown	0:Absent,1:Present within 4 weeks, 2:Persistent over 4 weeks, 9: Unknown
Respiratory failure	0:Absent,1:Present,9:Unknown	0:Absent,1:Present within 4 weeks, 2:Persistent over 4 weeks, 9: Unknown
6. Cardiovascular		
Loss of pulses	0:Absent,1:Present,9:Unknown	0:Absent,1:Present within 4 weeks, 2:Persistent over 4 weeks, 9: Unknown
Valvular heart disease	0:Absent,1:Present,9:Unknown	0:Absent,1:Present within 4 weeks, 2:Persistent over 4 weeks, 9: Unknown
Pericarditis	0:Absent,1:Present,9:Unknown	0:Absent,1:Present within 4 weeks, 2:Persistent over 4 weeks, 9: Unknown
Ischaemic cardiac pain	0:Absent,1:Present,9:Unknown	0:Absent,1:Present within 4 weeks, 2:Persistent over 4 weeks, 9: Unknown
Cardiomyopathy	0:Absent,1:Present,9:Unknown	0:Absent,1:Present within 4 weeks, 2:Persistent over 4 weeks, 9: Unknown
Congestive cardiac failure	0:Absent,1:Present,9:Unknown	0:Absent,1:Present within 4 weeks, 2:Persistent over 4 weeks, 9: Unknown
7. Abdominal		
Peritonitis	0:Absent,1:Present,9:Unknown	0:Absent,1:Present within 4 weeks, 2:Persistent over 4 weeks, 9: Unknown
Bloody diarrhoea	0:Absent,1:Present,9:Unknown	0:Absent,1:Present within 4 weeks, 2:Persistent over 4 weeks, 9: Unknown
Ischaemic abdominal pain	0:Absent,1:Present,9:Unknown	0:Absent,1:Present within 4 weeks, 2:Persistent over 4 weeks, 9: Unknown
8. Renal		
Hypertension (diastolic pressure >95 mmHg)	0:Absent,1:Present,9:Unknown	0:Absent,1:Present within 4 weeks, 2:Persistent over 4 weeks, 9: Unknown
Proteinuria >1+ or 0.2 g/day	0:Absent,1:Present,9:Unknown	0:Absent,1:Present within 4 weeks, 2:Persistent over 4 weeks, 9: Unknown
Haematuria ≥ 10 RBCs/hpf	0:Absent,1:Present,9:Unknown	0:Absent,1:Present within 4 weeks, 2:Persistent over 4 weeks, 9: Unknown
Serum creatinine > 1.4 mg/dl	0:Absent,1:Present,9:Unknown	0:Absent,1:Present within 4 weeks, 2:Persistent over 4 weeks, 9: Unknown
	serum Cr : mg/dl	serum Cr : mg/dll
Rise in serum creatinine >30% or fall in creatinine clearance >25%	0:Absent,1:Present,9:Unknown	0:Absent,1:Present within 4 weeks, 2:Persistent over 4 weeks, 9: Unknown
9. Nervous system		
Headache	0:Absent,1:Present,9:Unknown	0:Absent,1:Present within 4 weeks, 2:Persistent over 4 weeks, 9: Unknown
Meningitis	0:Absent,1:Present,9:Unknown	0:Absent,1:Present within 4 weeks, 2:Persistent over 4 weeks, 9: Unknown
Organic confusion	0:Absent,1:Present,9:Unknown	0:Absent,1:Present within 4 weeks, 2:Persistent over 4 weeks, 9: Unknown
Dementia	0:Absent,1:Present,9:Unknown	0:Absent,1:Present within 4 weeks, 2:Persistent over 4 weeks, 9: Unknown
Seizures	0:Absent,1:Present,9:Unknown	0:Absent,1:Present within 4 weeks, 2:Persistent over 4 weeks, 9: Unknown
Cerebrovascular accident	0:Absent,1:Present,9:Unknown	0:Absent,1:Present within 4 weeks, 2:Persistent over 4 weeks, 9: Unknown
Spinal cord lesion	0:Absent,1:Present,9:Unknown	0:Absent,1:Present within 4 weeks, 2:Persistent over 4 weeks, 9: Unknown

V. Laboratory findings	A. At definite diagnosis	B. At this screening
WBC count	_____/mm ³	_____/mm ³
Eosinophil count	_____% (or ____/mm ³)	_____% (or ____/mm ³)
Platlet count	____x 10 ⁴ /mm ³	____x 10 ⁴ /mm ³
IgE	____U/ ml	____U/ ml
MPO-ANCA (ELISA)	____EU/ml (MPO-ANCA normal range < ____)	____EU/ml (MPO-ANCA normal range < ____)
PR3-ANCA (ELISA)	____EU/ml (PR-3-ANCA normal range < ____)	____EU/ml (PR-3-ANCA normal range < ____)
p-ANCA (IF)	0:Positive,1:Negative,9:Unknown	0:Positive,1:Negative,9:Unknown
c-ANCA (IF)	0:Positive,1:Negative,9:Unknown	0:Positive,1:Negative,9:Unknown
Rheumatoid factor	0:Positive,1:Negative,9:Unknown	0:Positive,1:Negative,9:Unknown
If positive, specify the measurement method	1:Qualitative test 2:RAPA/RAHA ____times 3:Nephelometry ____IU/ml	1:Qualitative test 2:RAPA/RAHA ____times 3:Nephelometry ____IU/ml

VI. Treatment	C. At remission induction	B. At this screening
Corticosteroids If use, specify dosage convert into prednisolone	0:Not use, 1:Use, 9:Unknown ____mg/day	0:Not use, 1:Use, 9:Unknown ____mg/day
Cyclophosphamide	0:Not use, 1:Intravenous, 2:Per os, 9:Unknown	0:Not use, 1:Intravenous, 2:Per os, 9:Unknown
Other immunosuppressants If use, specify the drug	0:Not use, 1:Use, 9:Unknown	0:Not use, 1:Use, 9:Unknown
Intravenous immunoglobulin	0:Not use, 1:Use, 9:Unknown	0:Not use, 1:Use, 9:Unknown
Plasma exchange	0:Not use, 1:Use, 9:Unknown	0:Not use, 1:Use, 9:Unknown
Biologics If use, specify the drug	0:Not use, 1:Use, 9:Unknown	0:Not use, 1:Use, 9:Unknown

VII. Severity at this screening	
1. Irreversible damages due to vasculitis	0:Absent, 1:Present, 9:Unknown
If present, specify the damages	1:Renal failure (without hemodialysis), 2:Renal failure (with hemodialysis), 3:Mortor or sensory disturbance due to central nerve disorder, 4:Mortor or sensory disturbance due to peripheral nerve disorder, 5:Respiratory failure requires home oxygen therapy, 6:Heart failure more than NYHA III, 7:Tube feeding, 8:Others (____)