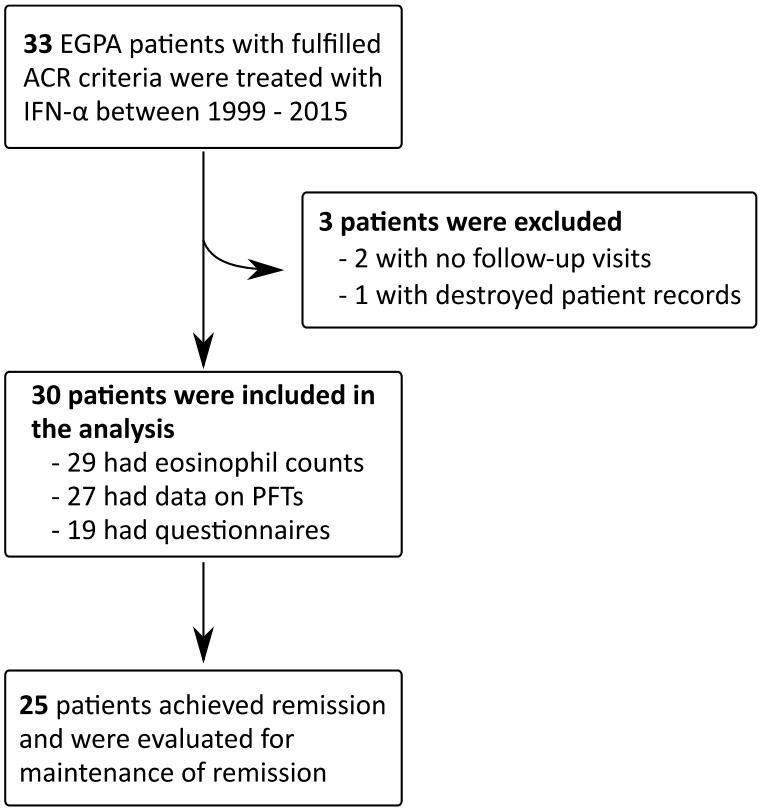
Online supplement to: Interferon-α for Induction and Maintenance of Remission in Eosinophilic Granulomatosis with Polyangiitis: A Single-center Retrospective Observational Cohort Study. *The Journal of Rheumatology.* doi:10.3899/jrheum.160907

ONLINE SUPPLEMENTARY CONTENT

Supplementary Figure 1. Study population.



EGPA: eosinophilic granulomatosis with polyangiitis; IFN: interferon; PFTs: pulmonary function tests.

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Supplementary Table 1. Biospy characteristics of 30 patients with EGPA treated with Interferon-α.

Characteristic, n (%)	**All (n=30)**
Biopsy performed	24 (80)
Biopsy findings	
Small- to medium-sized-vessel vasculitis	11 (37)
Extravascular eosinophils	22 (73)
Granuloma	5 (17)
Vasculitis with extravascular eosinophils	10 (67)
Granuloma with extravascular eosinophils	3 (10)
Loeffler Endocarditis	4 (13)
Biopsy sites	
Number of organs biopsied, median (Range)	1 (0-4)
Lung	19 (63)
Endomyocardium	5 (17)
Skin	4 (13)
Liver	2 (7)
Nasal sinuses	2 (10)
Gastrointestinal Tract	4 (13)
Muscle	1 (3)
Nerve	1 (3)
Kidney	1 (3)
Retroorbital mass	1 (3)

Supplementary Table 2. Features of cardiac involvement in 10 of 30 patients with EGPA treated with Interferon-α.

Clinical features, n (%)	**All (n=12)**
Acute myocardial ischemia	4
Myocarditis	4
Pericarditis	5
New arrhythmia	5
New valve regurgitation	2
Intracardial thrombus	1
Abnormal echocardiography*	9
Not performed	1
Abnormal cardiac magnetic resonance imaging	6
Not performed	6

* If pericardial effusion, ejection fraction <50%, wall motion abnormalities and or aortic or mitral regurgitation is present
† if additional edema or myocardial enhancement is present

Supplementary Table 3. Clinical surrogates for vasculitis (i.e. polyangiitis) in 19 of 30 patients with EGPA without histological evidence of vasculitis.

Clinical vasculitis surrogate, n (%)	**All (n=19)**
Mononeuritis simplex / multiplex*	5
Palpable purpura	2
Acute myocardial ischemia†	2
Episcleritis	1
Alveolar haemorrhage	1

* showing axonal damage on nerve conduction studies
† without evidence of coronary artery disease on cardiac catheterization

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Supplementary Table 4. Triggers for discontinuation of Interferon-α therapy in 23 / 30 patients with EGPA.

Trigger for discontinuation, n	All patients (n=23)
Lack of efficacy	5
No remission induction	1
Relapse*	4
Adverse events	13
Depression	4
Neuropathy	3
Autoimmune hepatitis	1
Liver damage	1
Anaemia	1
Alopecia	1
Nausea	1
Flushes	1
Pruritus	1
Discontinued whilst in remission†	3
Self-initiated discontinuation by the patient	1

At time of data collection, 7 patients were still receiving Interferon-α and 23 patients discontinued treatment.
* unresponsive to dose increase of interferon-α or prednisolone
† for de-escalation of therapy after long-term prednisolone-free remission by treating physician in (n=2) and by the patient because of scheduled surgery (n=1)

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Supplementary Table 5. Univariate analysis in outcomes (remission; relapse) by baseline characteristics.

		Achieved remission (25/30)		
Characteristics	n	yes	no	P*
Organ involvement				
ENT	22	19	3	0.383
Cardiac	10	7	3	0.139
Neurological	8	6	2	0.400
Skin	7	4	3	0.022
Eyes	2	2	0	0.520
GIT	2	2	0	0.520
Evidence of polyangiitis†	22	18	4	0.712
Relapsing EGPA	8	7	1	0.743
FFS (≥1)	8	4	4	0.003
BVAS at entry, median (IQR)	25	6 (4-10)	13.5 (6-17)	0.332
Age (years), median (IQR)	25	49 (43-55)	58 (51.5-63.5)	0.137

		Suffered relapse (16/25)		
Characteristics	n	yes	no	P*
Organ involvement				
ENT	19	13	6	0.058
Cardiac	7	5	2	0.658
Neurological	6	4	2	0.776
Skin	4	3	1	0.549
Eyes	2	2	0	0.549
GIT	2	0	2	0.058
Evidence of polyangiitis†	19	13	5	0.170
Relapsing EGPA	7	5	2	0.525
FFS (≥1)	4	1	3	0.091
BVAS at entry, median (IQR)	21	6 (4-12)	5 (2.5-9)	0.257
Age (years), median (IQR)	21	50 (44-55)	48 (42.5-55)	0.944

FFS: Five-Factor Score; ENT: Ear Nose Throat; GIT: Gastrointestinal-Tract; BVAS: Birmingham Vasculitis Activity Score

*group comparison using χ^2.

† defined as presence of necrotizing vasculitis on biopsy or strong clinical surrogate of polyangiitis (myocardial ischemia due to coronaritis; palpable purpura; episcleritis; mononeuritis multiplex, alveolar hemorrhage).