

Supplementary Table S1: Clinical features at presentation[§]

	Italy (n=127)	India (n=191)	Odd's ratio (95% CI)	p value* corrected for (Italy vs multiple India) testing
Clinical features at presentation [n(%)]				
None	4 (3)	0 (0)	-	0.451 ^b
Seizure	0 (0)	9 (5)	-	0.266 ^b
Documented TAK retinopathy	0 (0)	2 (1)	-	>0.999 ^b
Aortic regurgitation	0 (0)	11 (6)	-	0.090 ^b
Acute coronary syndrome	7 (5)	0 (0)	-	0.035^b
Other	17 (13)	22 (12)	-	-
Specify list with numbers	Dyspnoea (n=9) Flank pain (n=1) Cardiac arrest (n=1) Myocarditis (n=1) Lung infarction (n=2) Pulmonary arterial hypertension (n=1) Digital necrosis (n=1)	Palpitations (n=8) Digital necrosis (n=1) Hemoptysis (n=2) Breathlessness (n=9) Oral ulcers (n=1) Occasional upper limb numbness (n=1)		
Baseline investigations				
ESR (mm/hr) [mean (± SD)]	61 ± 32	46 ± 32 (n=186)	-	<0.001
CRP (mg/L) [mean (± SD)]	37 ± 42	24 ± 32 (n=179)	-	0.007
Hemoglobin (g/dL) [mean (± SD)]	11.8 ± 1.2	11.4 ± 2.0 (n=185)	-	0.128

^{\$}Those clinical features other than the ones depicted in Figure 1 are presented here.

* Chi squared^a/ Fisher's exact^b for proportions

BP – Blood pressure; 95% CI – 95% confidence interval; CRP – C-reactive protein; ESR – Erythrocyte sedimentation rate; SD – Standard deviation; TAK – Takayasu arteritis; TIA – Transient ischemic attack.

NA – Not assessable

p values <0.05 are highlighted in bold

Supplementary Table S2: Imaging modalities and vascular involvement at initial assessment

	Italy [n(%)]	India [n(%)]	Odd's ratio (95% CI) (Italy vs India)	p value corrected for multiple testing
Imaging modalities\$	(n=127)	(n=191)		
PET-MRI	1 (0.8)	0 (0)	-	0.953 ^b
Vascular involvement	n=127	n=190		
Coronary	15 (12)	3 (2)	8.35 (2.49 – 27.52)	0.005^b
Right subclavian	65 (51)	75 (39)	1.61 (1.02 – 2.55)	0.590 ^a
Left subclavian	81 (64)	138 (73)	0.66 (0.41 – 1.08)	0.888 ^a
Right carotid	74 (58)	75 (39)	2.14 (1.36 – 3.34)	0.022^a
Left carotid	79 (62)	99 (52)	1.51 (0.96 – 2.36)	0.823 ^a
Right vertebral	22 (17)	6 (3)	6.43 (2.52 – 15.55)	<0.001 ^a
Left vertebral	18 (14)	22 (12)	1.26 (0.64 – 2.38)	>0.999 ^a
Pulmonary	21 (17)	11 (6)	3.22 (1.46 – 7.21)	0.040^a
Brachiocephalic	34 (27)	48 (25)	1.08 (0.64 – 1.79)	>0.999 ^a
Ascending aorta	36 (28)	36 (19)	1.69 (1.01 – 2.84)	0.679 ^a
Arch of aorta	44 (35)	67 (35)	0.97 (0.61 – 1.54)	>0.999 ^a
Descending thoracic aorta	45 (35)	94 (49)	0.56 (0.36 – 0.89)	0.259 ^a
Abdominal aorta	57 (45)	105 (55)	0.66 (0.42 – 1.03)	0.797 ^a
Celiac trunk	38 (30)	49 (26)	1.23 (0.75 – 1.99)	>0.999 ^a
Superior mesenteric artery	36 (28)	44 (23)	1.31 (0.78 – 2.19)	>0.999 ^a

Inferior mesenteric artery	4 (3)	8 (4)	0.74 (0.24 – 2.40)	>0.999 ^b
Right renal	27 (21)	81 (43)	0.36 (0.22 – 0.61)	0.002^a
Left renal	25 (20)	78 (41)	0.35 (0.21 – 0.60)	0.002^a
Right iliac	21 (16)	12 (6)	2.94 (1.37 – 6.15)	0.074 ^a
Left iliac	21 (16)	13 (7)	2.70 (1.29 – 5.37)	0.129 ^a
Right femoral	4 (3)	2 (1)	3.06 (0.70 – 16.21)	0.996 ^b
Left femoral	2 (2)	3 (2)	0.99 (0.18 – 4.94)	>0.999 ^b

^{\$}Those imaging modalities other than the ones depicted in Figure 2A are presented here.

* Chi squared^a/ Fisher's exact^b for proportions

95% CI – 95% confidence interval; C+ - Coronary involvement; CT – Computerized tomography; PET – Positron emission tomography; MRI – Magnetic resonance imaging; NA – Not assessable; P+ - Pulmonary involvement
p values <0.05 are highlighted in bold

Supplementary Table S3: Treatments received

	Italy (n=127)	India (n=191)	p value*
Glucocorticoids			
n(%)	124 (98)	148 (77)	<0.001^b
on intravenous methylprednisolone	7 (5)	2 (1)	0.033^b
n(%)			
Starting dose (mean with SD)	48.1 ± 13.3	33.2 ± 14.6 (n=143)	<0.001
Continuing at last follow-up	88 (71)	118 (80)	0.093 ^a
n(%)			
Duration in months (mean ± SD)	103 ± 90	38 ± 36 (n=140)	<0.001
Percentage reduction in prednisolone at last visit	90 ± 14	87 ± 17 (n=119)	0.134
Methotrexate			
n(%)	102 (80)	80 (42)	<0.001^a
Continuing at last follow-up	77 (75)	40 (50%)	<0.001^a
n(%)			
Duration in months (mean ± SD)	57 ± 58	33 ± 32 (n=77)	0.001
Leflunomide			
n(%)	12 (9)	2 (1)	<0.001^b
Continuing at last follow-up	11 (92)	1 (50%)	0.275 ^b
n(%)			
Duration in months (mean ± SD)	13 ± 7	8 ± 7 (n=2)	0.368
Azathioprine			
n(%)	42 (33)	28 (15)	<0.001^a
Continuing at last follow-up	11 (26)	9 (32)	0.589 ^a
n(%)			
Duration in months (mean ± SD)	55 ± 55	31 ± 34 (n=26)	0.050
Mycophenolate			
n (%)	24 (19)	34 (18)	0.804 ^a
Continuing at last follow-up	9 (38)	19 (56)	0.168 ^a
n(%)			
Duration in months (mean ± SD)	31 ± 37	17 ± 17 (n=34)	0.057
Cyclosporine A			
n(%)	3 (2)	0	0.063 ^b

Continuing at last follow-up n(%)	1 (33)	0	-
Duration in months (mean ± SD)	26 (19)	0	-
Sirolimus			
n(%)	5 (4)	0	0.001 ^b
Continuing at last follow-up n(%)	2 (40)	0	-
Duration in months (mean ± SD)	80 (64)	0	-
Tacrolimus			
n(%)	1 (0.8)	68 (36)	<0.001 ^b
Continuing at last follow-up n(%)	0 (0)	54 (79)	0.217 ^b
Duration in months (mean ± SD)	22	18 ± 20 (n=68)	-
Cyclophosphamide			
n(%)	0	4 (2)	0.153
Continuing at last follow-up n(%)	0	0 (0)	-
Duration in months (mean ± SD)	0	6 ± 2 (n=4)	-
Total number of csDMARDs received (mean with SD)			
Infliximab			
n(%)	58 (46)	0	<0.001 ^b
Continuing at last follow-up n(%)	40 (69)	0	-
Duration in months (mean ± SD)	58 ± 51	0	
Adalimumab			
n(%)	22 (17)	1 (0.5)	<0.001 ^b
Continuing at last follow-up n(%)	12 (55)	1 (100%)	0.999 ^b
Duration in months (mean ± SD)	32 ± 31	10	-
Golimumab			
n(%)	13 (10)	0	<0.001 ^b
Continuing at last follow-up n(%)	7 (54)	0	-
Duration in months (mean ± SD)	21 ± 21	0	-

Certolizumab pegol			
n(%)	1 (0.8)	0	0.399 ^b
Continuing at last follow-up	0 (0)	0	-
n(%)			
Duration in months (mean ± SD)	5	0	-
Etanercept			
n(%)	2 (2)	0	0.159 ^b
Continuing at last follow-up	0 (0)	0	-
n(%)			
Duration in months (mean ± SD)	14 ± 3	0	-
Tocilizumab			
n(%)	22 (17)	4 (2)	<0.001 ^b
Continuing at last follow-up	11 (50)	0 (0)	0.113 ^b
n(%)			
Duration in months (mean ± SD)	30 ± 32	10 ± 11 (n=4)	0.235
Abatacept			
n(%)	1 (0.8)	0	0.399 ^b
Continuing at last follow-up	0 (0)	0	-
n(%)			
Duration in months (mean ± SD)	19	0	-
Rituximab			
n(%)	1 (0.8)	0	0.399 ^b
Continuing at last follow-up	0 (0)	0	-
n(%)			
Duration in months (mean ± SD)	3	0	-
Anakinra			
n(%)	3 (2)	0	0.063 ^b
Continuing at last follow-up	1 (33)	0	-
n(%)			
Duration in months (mean ± SD)	11 ± 3	0	-
Tofacitinib			
n(%)	3 (2)	1 (0.5)	0.306 ^b
Continuing at last follow-up	3 (100)	1 (100%)	>0.999 ^b
n(%)			
Duration in months (mean ± SD)	28 ± 9	12	-

Baricitinib			
n(%)	0	0	-
Continuing at last follow-up	0	0	-
n(%)			
Duration in months (mean ± SD)	0	0	-
Total number of ts or bDMARDs received (mean with SD)	1 ± 1.2	0.04 ± 0.21	<0.001
Antihypertensives			
n(%)	56 (44)	148 (77)	<0.001^a
Mean (± SD) number of antihypertensives at presentation	0.9 ± 1.2	1.7 ± 1.4	<0.001
Aspirin	71 (56)	50 (26)	<0.001^a
Clopidogrel	9 (7)	21 (11)	0.243
Statin	43 (34)	23 (12)	<0.001^a

* Chi squared^a/ Fisher's exact^b for proportions

SD – Standard deviation

p values <0.05 are highlighted in bold

Supplementary Table S4: Reasons for failure of disease-modifying anti-rheumatic drugs

Reason for failure	Italy (n=95)	India (n=66)	p value* corrected for multiple testing
Switch to another DMARD	26 (27%)	31 (47%)	0.031
Suspension of DMARD	12 (13%)	19 (29%)	0.031
Add-on of another DMARD	57 (60%)	16 (24%)	<0.001
DMARD – Disease-modifying anti-rheumatic drug			
*Chi square test			
p values <0.05 are highlighted in bold			

Supplementary Table S5: Choice of first and second-line disease-modifying anti-rheumatic drugs

	Italy (n=114)	India (n=143)
First-line DMARD	n= 114	n=143
Methotrexate (85)		Methotrexate (64)
Azathioprine (20)		Tacrolimus (50)
Leflunomide (1)		Azathioprine (12)
Mycophenolate (6)		Mycophenolate (12)
Infliximab (2)		Cyclophosphamide followed by Azathioprine (2)
		Cyclophosphamide (1)
		Tocilizumab (1)
		Methotrexate + Mycophenolate (1)
Second-line DMARD (switch or add-on)	n=81	n=50
Infliximab (41)		Mycophenolate (15)
Azathioprine (9)		Azathioprine (11)
Methotrexate (8)		Methotrexate (10)
Golimumab (6)		Tacrolimus (8)
Adalimumab (6)		Leflunomide (2)
Tocilizumab (5)		Tocilizumab (2)
Leflunomide (2)		Adalimumab (1)
Sirolimus (2)		Cyclophosphamide (1)
Tofacitinib (1)		
Mycophenolate (1)		
<u>DMARD – Disease-modifying anti-rheumatic drug</u>		