

Gen Amir Chand RAE in field of non communicable diseases
Exciting world of Vasculitis: Care and Clinical Research



Aman Sharma

Professor

Clinical Immunology & Rheumatology Wing

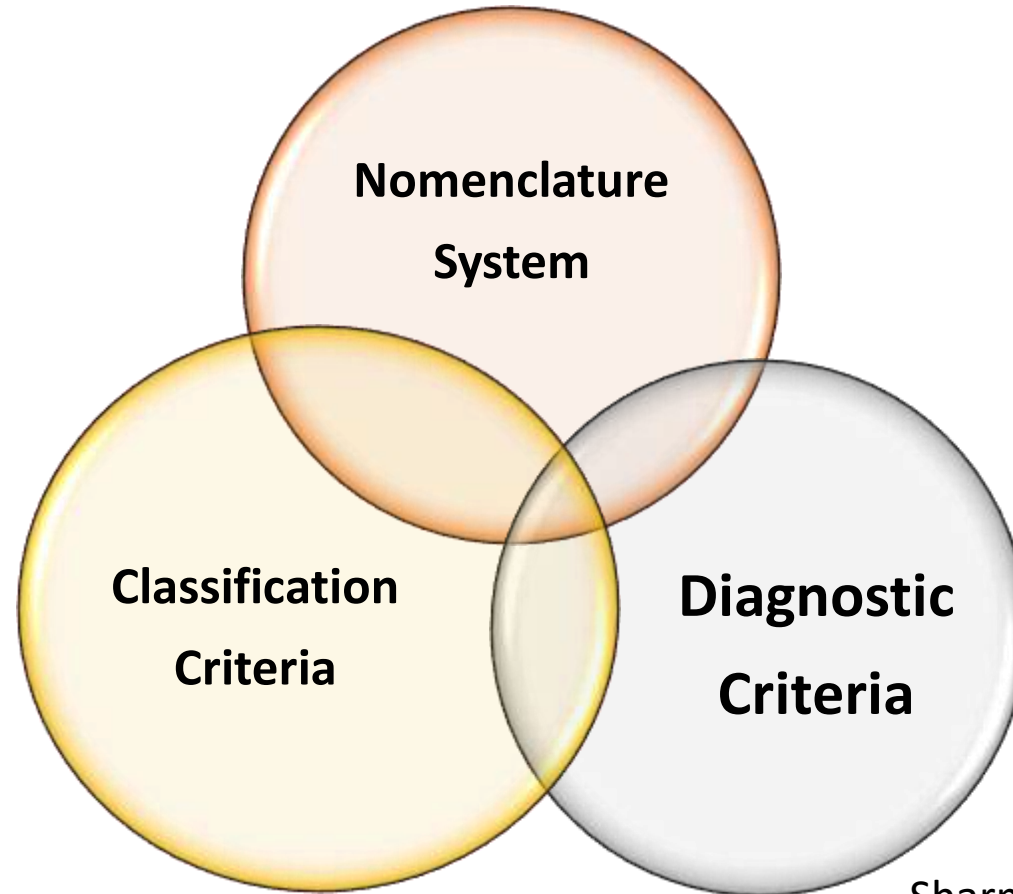
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Nomenclature, classification and diagnostic criteria in systemic vasculitis – ‘A work in progress’



Sharma A. Ind J Rheumatol. 2013; 8:99-101.



Validation of the consensus methodology algorithm for the classification of systemic necrotizing vasculitis in Indian patients

Aman SHARMA,¹ Tarun MITTAL,² Roopa RAJAN,³ Manish RATHI,² Ritambhra NADA,⁴ Ranjana W. MINZ,⁵ Kusum JOSHI,⁴ Vinay SAKHUJA,² and Surjit SINGH¹

Table 1 Reclassification of patients with systemic necrotizing vasculitis using Watts' algorithm

79 Patients 2007-11

	No. of patients (% of all patients) as per initial diagnosis	No. of patients reclassified in the same diagnostic subcategory (%)	
Present study			
EGPA	4 (5.1)	4 (100)	One patient reclassified using Lanham criteria
GPA	45 (57.7)	45 (100)	42 patients – 2a; two patients – 2c; one patient 2d
MPA	4 (5.1)	4 (100)	One patient excluded from analyses
PAN	18 (23.1)	13 (72.2)	Two patients reclassified to MPA 3a, three patients reclassified to unclassifiable
Unclassifiable	7 (9.0)	0 (0)	One patient reclassified to GPA, six patients reclassified to MPA 3a
Total	78 (100)	66 (84.6)	

EGPA, eosinophilic granulomatosis with polyangiitis; GPA, granulomatosis with polyangiitis; MPA, microscopic polyangiitis; PAN, polyarteritis nodosa.

Sharma A. Int J Rheum Dis. 2013



2022 American College of Rheumatology/European Alliance of Associations for Rheumatology classification criteria for granulomatosis with polyangiitis

2022 American College of Rheumatology/European Alliance of Associations for Rheumatology Classification Criteria for Eosinophilic Granulomatosis with Polyangiitis

2022 American College of Rheumatology/European Alliance of Associations for Rheumatology classification criteria for microscopic polyangiitis



ACR/EULAR 2017 provisional classification criteria of Granulomatosis with polyangiitis(GPA)

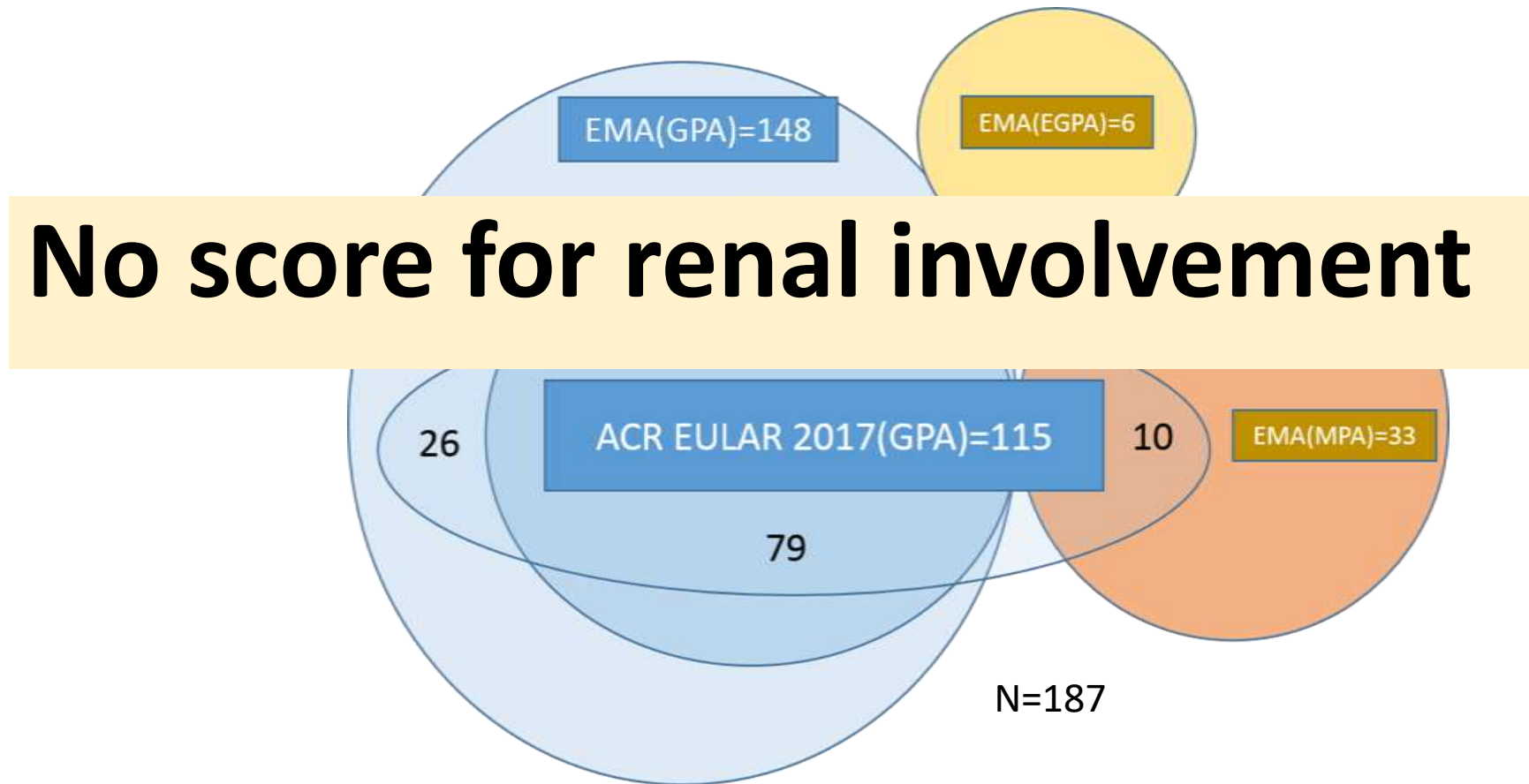
This criteria set is use when diagnosis of small or medium vessel vasculitis has been made, in order to classify the patient as having GPA

Clinical	Bloody nasal discharge, ulcers, crusting or sino-nasal congestion	3
	Nasal polyps	-4
	Hearing loss or reduction	1
	Cartilage involvement	2
Tests	Red or painful eyes	1
	cANCA or PR3 antibody	5
	Eosinophil count $\geq 1(x10^9/L)$	-3
	Nodules, mass, or cavity on chest imaging	2
	Granuloma on biopsy	3

Add scores for 9 items; **a score of ≥ 5** is needed for classification of GPA



Validation of the ACR EULAR provisional 2017 classification criteria of GPA amongst Indian patients with AAV (n=187)



Sharma A et al Arthritis Rheumatol. 2017; 69 (suppl 10).



2022 AMERICAN COLLEGE OF RHEUMATOLOGY / EUROPEAN ALLIANCE OF ASSOCIATIONS FOR RHEUMATOLOGY

CLASSIFICATION CRITERIA FOR **GRANULOMATOSIS WITH POLYANGIITIS**

CONSIDERATIONS WHEN APPLYING THESE CRITERIA

- These classification criteria should be applied to classify a patient as having granulomatosis with polyangiitis when a diagnosis of small- or medium-vessel vasculitis has been made
- Alternate diagnoses mimicking vasculitis should be excluded prior to applying the criteria

CLINICAL CRITERIA

Nasal involvement: bloody discharge, ulcers, crusting, congestion, blockage, or septal defect / perforation	+3
Cartilaginous involvement (inflammation of ear or nose cartilage, hoarse voice or stridor, endobronchial involvement, or saddle nose deformity)	+2
Conductive or sensorineural hearing loss	+1

LABORATORY, IMAGING, AND BIOPSY CRITERIA

Positive test for cytoplasmic antineutrophil cytoplasmic antibodies (cANCA) or antiproteinase 3 (anti-PR3) antibodies	+5
Pulmonary nodules, mass, or cavitation on chest imaging	+2
Granuloma, extravascular granulomatous inflammation, or giant cells on biopsy	+2
Inflammation, consolidation, or effusion of the nasal/paranasal sinuses, or mastoiditis on imaging	+1
Pauci-immune glomerulonephritis on biopsy	+1
Positive test for perinuclear antineutrophil cytoplasmic antibodies (pANCA) or antityeloperoxidase (anti-MPO) antibodies	-1
Blood eosinophil count $\geq 1 \times 10^9$ /liter	-4



Validation of the provisional seven-item criteria for the diagnosis of polyarteritis nodosa


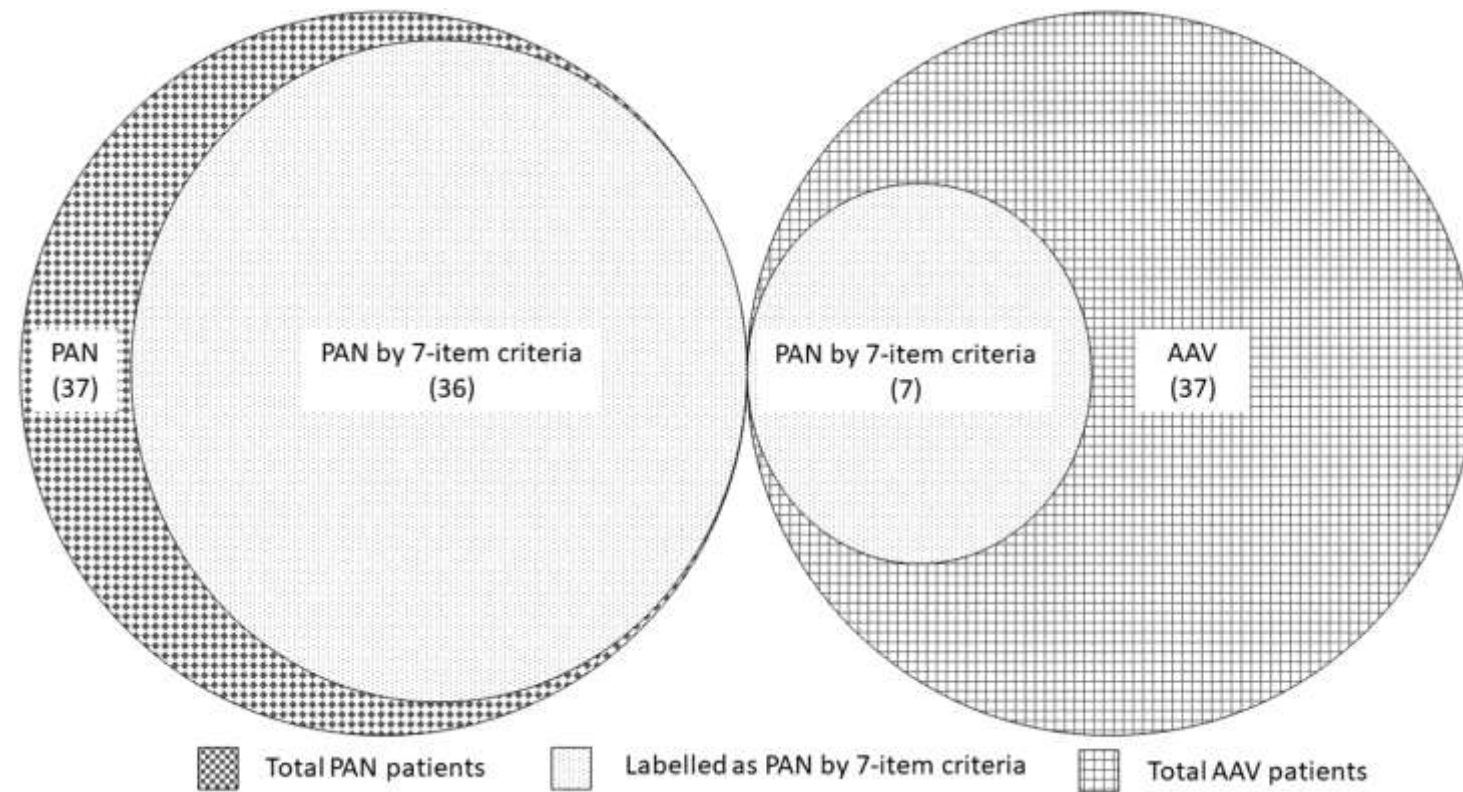
GSRSNK Naidu¹ · Chirag Rajkumar Kopp¹ · Vikas Sharma² · Manphool Singhal³ · Benzeeta Pinto⁴ · Varun Dhir¹ · Ritambhra Nada⁵ · Ranjana W. Minz⁶ · Sanjay Jain¹ · Aman Sharma¹ 

Fig. 1 Diagnostic label of PAN and AAV patients as per the seven-item criteria. AAV ANCA associated vasculitis, PAN Polyarteritis Nodosa



Rheumatology International (2021) 41:1651–1655



Chapel Hill 2012

- **Large Vessel Vasculitis (LVV)**

- Takayasu Arteritis (TAK)
- Giant Cell Arteritis (GCA)

- **Medium Vessel Vasculitis (MVV)**

- Polyarteritis Nodosa (PAN)
- Kawasaki Disease (KD)

- **Small Vessel Vasculitis (SVV)**

- **ANCA Associated Vasculitis**
 - Microscopic Polyangiitis (MPA)
 - Granulomatosis with Polyangiitis (GPA)
 - Eosinophilic Granulomatosis with Polyangiitis (EGPA)
- Immune complex SVV

- **Variable Vessel Vasculitis (VVV)**

- Behçet Disease (BD)
- Cogan's Syndrome

- **Single Organ Vasculitis (SOV)**

- Cutaneous Leukocytoclastic Angiitis
- Cutaneous Arteritis
- Primary Angiitis of the CNS (PACNS)
- Isolated Aortitis

- **Vasculitis Associated with Systemic Disease**

- Lupus vasculitis
- Rheumatoid vasculitis
- Sarcoid vasculitis

- **Vasculitis with Probable Etiology**

- HBV/HCV-associated vasculitis
- Drug-associated immune complex vasculitis
- Drug-associated ANCA-associated Vasculitis
- Cancer associated vasculitis



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Giant cell arteritis: Its ophthalmic manifestations

Sohan Singh Hayreh

South India^[30-35], and Assam.^[36] I am from Punjab originally, and I developed GCA in 2011, and two of my siblings also developed it at other times. It seems GCA is underdiagnosed in India. This may be because of the prevalent impression among Indian physicians that **GCA does not exist in India.**

Indian J Ophthalmol 2021;69:227-35.



Giant cell arteritis in India: Report from a tertiary care center along with total published experience from India

Aman Sharma, Vinay Sagar, Mahesh Prakash¹, Vikas Gupta², Niranajan Khaire, Benzeeta Pinto, Varun Dhir, Amanjit Bal³, Ashish Aggarwal, Susheel Kumar, Kusum Sharma⁴, Manish Rathi⁵, Ashim Das³, Ramandeep Singh⁶, Surjit Singh, Amod Gupta⁶

	Published Indian series and case reports						Present series (Northern India)	Cumulative Indian data
	Western India ^[5]	Eastern India ^[8]	Western India ^[6]	Southern India ^[9]	Southern India ^[7]	Indian case reports ^[10-13]		
Number of cases	7	4	16	9	15	4	17	72
Age (years)	66.8 (58-73)	74.5	66.5 (58-78)	70 (59-81)	67.53 (52-81)	66.75 (63-74)	67 (54-82)	67.77
Sex ratio (male:female)	1:1.3	1:1	1:1	1.25:1	1.5:1	1:1	2.4:1	1.32:1
Delay in diagnosis (days)	345	188	155 (15-720)	52 (7-140)	77 (20-120)	60.5 (7-90)	53 (4-180)	116.85
Headache (%)	7 (100)	4 (100)	15 (93.7)	6 (66.7)	15 (100)	4 (100)	17 (100)	68 (94.4)
Fever (%)	2 (28.6)	00	9 (56.2)	4 (44.4)	00	3 (75)	3 (17.6)	21 (29)
Malaise (%)	1	2 (50)	NA	NA	NA	1 (25)	5 (29.4)	9 (12.5)
Polymyalgia rheumatica (%)	1 (14.2)	3 (75)	5 (31.2)	1 (11.1)	8 (53.3)	0 (0)	7 (41)	5 (34.7)
Vision loss (%)	2 (28.6)	1 (25)	3 (2-blurring,	2 (blurring)	3 (all	2 (1-complete	10 (5	23 (32)



Giant cell arteritis can occur in people of colour

Tiara Gill, Michael Putman, Sebastian E Sattui,
Shahir Hamdulay, Richard Conway, David F L Liew,
Aman Sharma, John H Stone, Sarah L Mackie, *Puja Mehta

Country	Number of patients diagnosed with GCA (n)	Study Design	Population ethnicity (%)	GCA diagnostic criteria	Positive temporal artery biopsy (%)	Reference
USA Baltimore	92	Retrospective cohort, multicentre	White (82) Black (15)	TAB	100	¹ Gruener 2019
USA Multicentre	32	Retrospective cohort	African Caribbean (100)	TAB	100	² Garrity 2012
USA Washington	50	Retrospective cohort	African -American (100)	TAB	100	³ Gilbert 1999
India Multicentre	72	Case series	Indian (100)	ACR 1990 ⁹	64 (46/72)	⁴ Sharma 2015
Japan	19	Retrospective cohort	Japanese (100)	ACR 1990	73 (11/15)	⁵ Imai 2011
Singapore	7	Case series	Chinese (100)	TAB	100	⁶ Cullen 2010
China	70	Retrospective cohort	Chinese (100)	ACR 1990 (96%; 67/70) PET (4%; 3/70)	71.4 (42/70)	⁷ Sun 2016
UK London	83	Retrospective cohort	White (49) Non-white (51)	ACR 1990 (60%; 50/83) TAB (39%; 33/83)	39 (33/83)	Personal communication from author (SH)

www.thelancet.com/rheumatology Vol 5 April 2023



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ORIGINAL ARTICLE

F-18 FDG PET/CT in the evaluation of Takayasu arteritis: An experience from the tropics

Sampath Santhosh, MD,^a Bhagwant Rai Mittal, MD, DNB,^a
Shankaramurthy Gayana, MD,^a Anish Bhattacharya, DNB,^a Aman Sharma, MD,^b

PET/CT in detecting active disease

- **Sensitivity- 83.3%**
- **Specificity- 90%**
- **PPV-77%**
- **NPV-93.3%**

J Nuclear Cardiol 2014

amansharma74@yahoo.com, @AmansharmPGI



Corticosteroid monotherapy for the management of Takayasu arteritis—a systematic review and meta-analysis

Durga Prasanna Misra¹ · Upendra Rathore¹ · Pallavi Patro² · Vikas Agarwal¹ · Aman Sharma³

A systematic review of clinical and preclinical evidences for Janus kinase inhibitors in large vessel vasculitis

Upendra Rathore¹ · Darpan Radheshyam Thakare¹ · Pallavi Patro² · Vikas Agarwal¹ · Aman Sharma³
Durga Prasanna Misra¹

Patient-Reported Outcome Measures in Takayasu Arteritis: A Systematic Review and Meta-Analysis

Durga P. Misra · Upendra Rathore · Pallavi Patro ·
Vikas Agarwal · Aman Sharma

Highly cited papers in Takayasu arteritis on Web of Science and Scopus: cross-sectional analysis

Durga Prasanna Misra¹ · Vikas Agarwal¹ · Armen Yuri Gasparyan² · Olena Zimba³ · Aman Sharma⁴



Phenotype to genotype approach

V

Vacuoles

E

E enzyme

X

X linked

A

Autoinflammatory

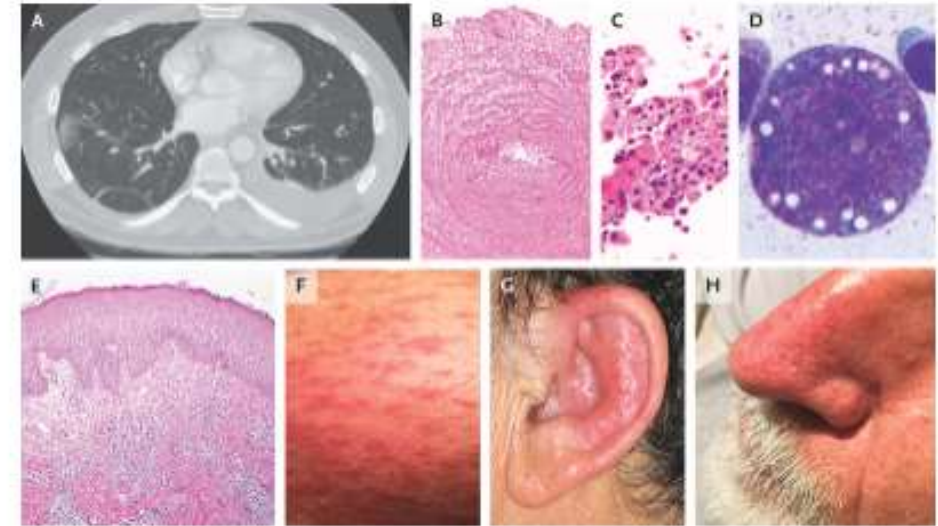
S

Somatic

The NEW ENGLAND JOURNAL of MEDICINE

ORIGINAL ARTICLE

Somatic Mutations in *UBA1* and Severe Adult-Onset Autoinflammatory Disease



Clinical Manifestations of the VEXAS Syndrome.

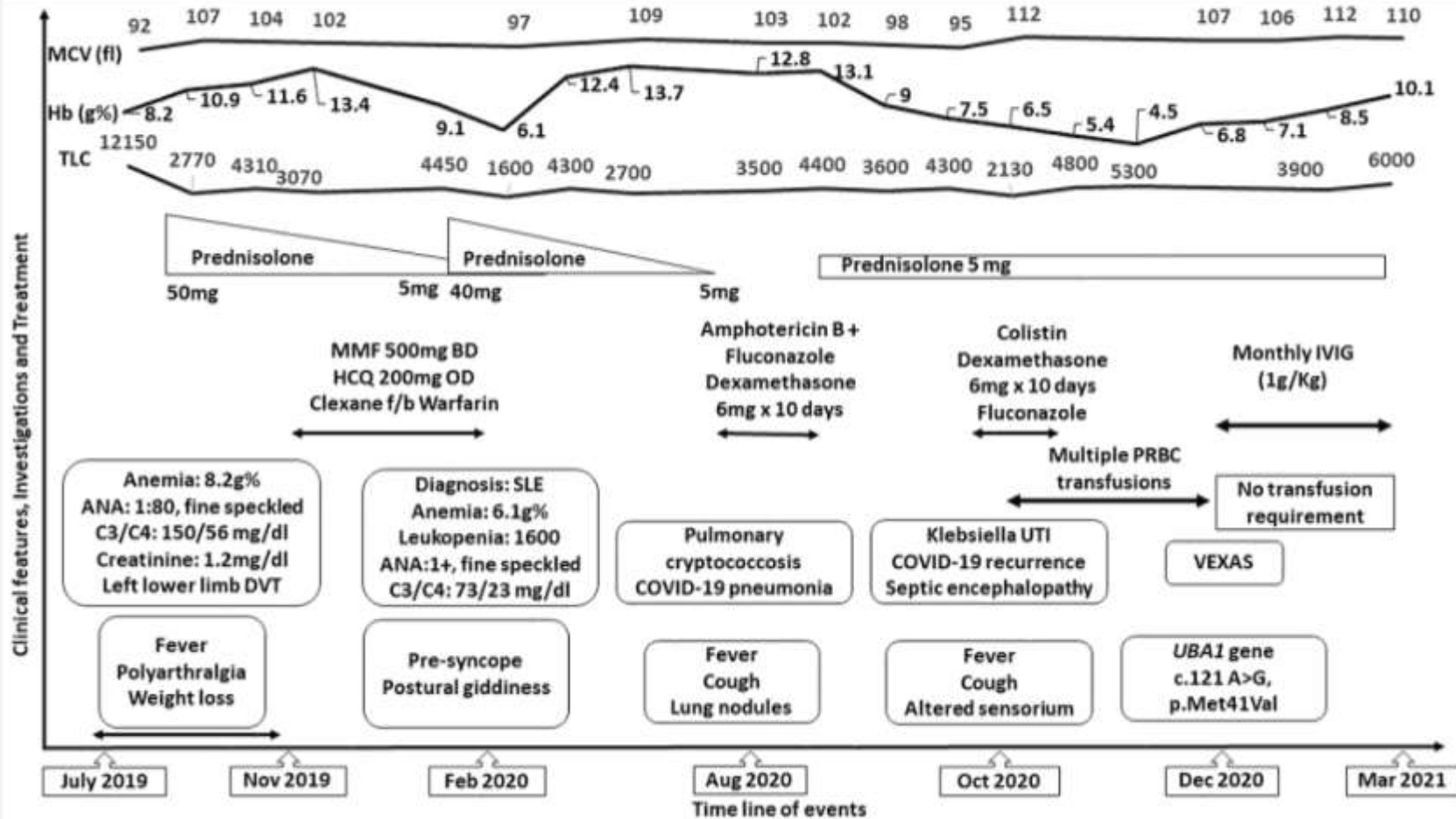
N Engl J Med. 2020;383(27):2628-38



Title: VEXAS syndrome with systemic lupus erythematosus- expanding the spectrum of associated conditions.

Keywords: VEXAS syndrome, Systemic lupus erythematosus.

Authors: Aman Sharma¹ M.D. FRCP(London), GSRSNK Naidu¹ D.M., Prateek Deo¹ M.D., David B. Beck² M.D., PhD.



Arthritis & Rheumatology, Volume: 74, Issue: 2, Pages: 369-371, First published: 30 August 2021,



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 - Eosinophilic Granulomatosis with Polyangiitis (EGPA)
 - Immune complex SVV
 - Anti-GBM disease
 - Cryoglobulinemic Vasculitis
 - IgA Vasculitis (HSP)
 - Hypocomplementemic Urticarial Vasculitis
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 - Drug-associated ANCA-associated Vasculitis
 - Cancer associated vasculitis

Arthritis Rheum 2013; 65: 1-11





ORIGINAL ARTICLE

Polyarteritis nodosa in north India: clinical manifestations and outcomes

Aman SHARMA,¹ Benzeeta PINTO,¹ Aadhaar DHOORIA,¹ Manish RATHI,²
Manphool SINGHAL,³ Varun DHIR,¹ Kusum SHARMA,⁴ Mahesh PARKASH,³
Manish MODI,⁵ Rajesh VIJAYVERGIYA,⁶ Saroj K. SINHA,⁷ Ritambhra NADA,⁸
Ranjana Walkar MINZ⁹ and Surjit SINGH¹

Departments of ¹Internal Medicine, ²Nephrology, ³Radiodiagnosis, ⁴Medical Microbiology, ⁵Neurology, ⁶Cardiology, ⁷Gastroenterology, ⁸Histopathology, and ⁹Department of Immunopathology, Post Graduate Institute of Medical Education and Research, Chandigarh, India

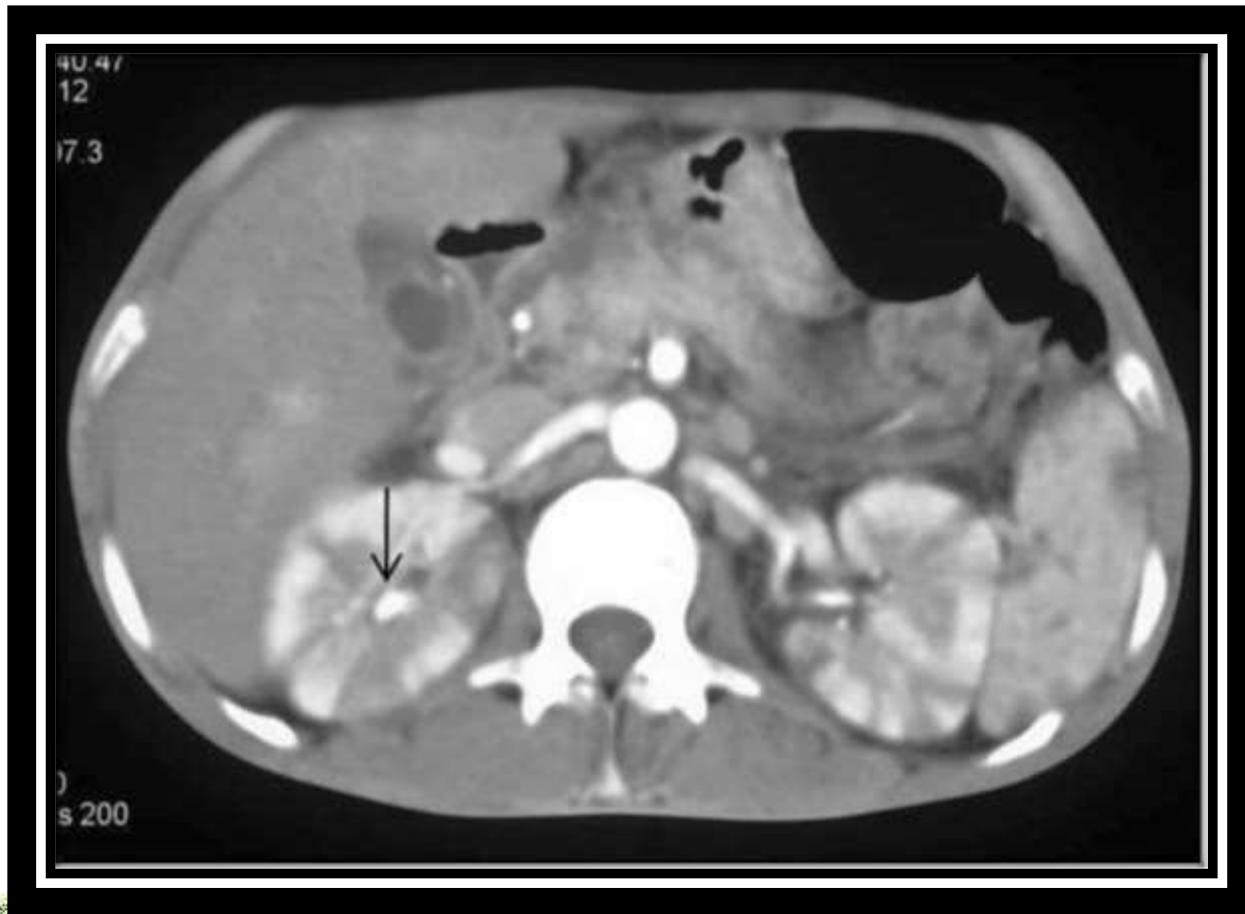


Characteristic	Overall (<i>n</i> = 27)	Non HBV related (<i>n</i> = 20)	HBV related (<i>n</i> = 7)	<i>P</i> -valu
Age	38.4 ± 13.2	38.25 ± 14.25	39.0 ± 10.53	0.900
Sex (M:F)	20 : 7	13 : 7	7 : 0	0.069
Constitutional symptoms				
Fever	14 (51.9%)	10 (50%)	4 (57.1%)	0.745
Weight loss	20 (74.1%)	13 (65%)	7 (100%)	0.069
Musculoskeletal symptoms	10 (37.0%)	8 (40.0%)	2 (28.6%)	0.590
Cutaneous involvement	9 (33.3%)	8 (40%)	1 (14.3%)	0.214
Testicular pain (male = 20)	6 (30.0%)	4 (30.8%)	2 (28.6%)	0.919
Peripheral gangrene	8 (29.6%)	7 (35.0%)	1 (14.3%)	0.302
Renal involvement	16 (59.3%)	10 (50%)	6 (85.7%)	0.098
Accelerated hypertension	11 (40.7%)	7 (35.0%)	4 (57.1%)	0.305
Proteinuria	10 (37.0%)	6 (30%)	4 (57.1%)	0.790
Elevated creatinine	6 (22.2%)	5 (25%)	1 (14.3%)	0.182
Hematuria	7 (25.9%)	6 (30%)	1 (14.3%)	0.091
Gastrointestinal involvement	8 (29.6%)	8 (40%)	0	0.046
Ischemic abdominal pain	6 (22.2%)	6 (30%)	0	0.100
Gastrointestinal bleed	4 (14.8%)	4 (20%)	0	0.200
Surgery	2 (7.4%)	2 (10.0%)	0	0.385
Neurological involvement	24 (88.9%)	17 (85%)	7 (100%)	0.277
Peripheral nervous system	22 (81.5%)	15 (75%)	7 (100%)	0.143
Central nervous system	4 (14.8%)	4 (20%)	0	0.200
Cardiac involvement (myocarditis)	4 (14.8%)	3 (15.0%)	1 (14.3%)	0.963



Role of multidetector abdominal CT in the evaluation of abnormalities in polyarteritis nodosa

M. Singhal^{a,*}, P. Gupta^a, A. Sharma^b, A. Lal^a, M. Rathi^c, N. Khandelwal^a



Clin Radiol 2015

amansharma74@yahoo.com, @AmansharmPGI



ORIGINAL ARTICLE

Early-Onset Stroke and Vasculopathy Associated with Mutations in ADA2

Q. Zhou et al, N Engl J Med 370;10: 2014

The NEW ENGLAND JOURNAL of MEDICINE

ORIGINAL ARTICLE










Mutant Adenosine Deaminase 2 in a Polyarteritis Nodosa Vasculopathy

Paulina Navon Elkan et al. N Engl J Med 370;10: 2014

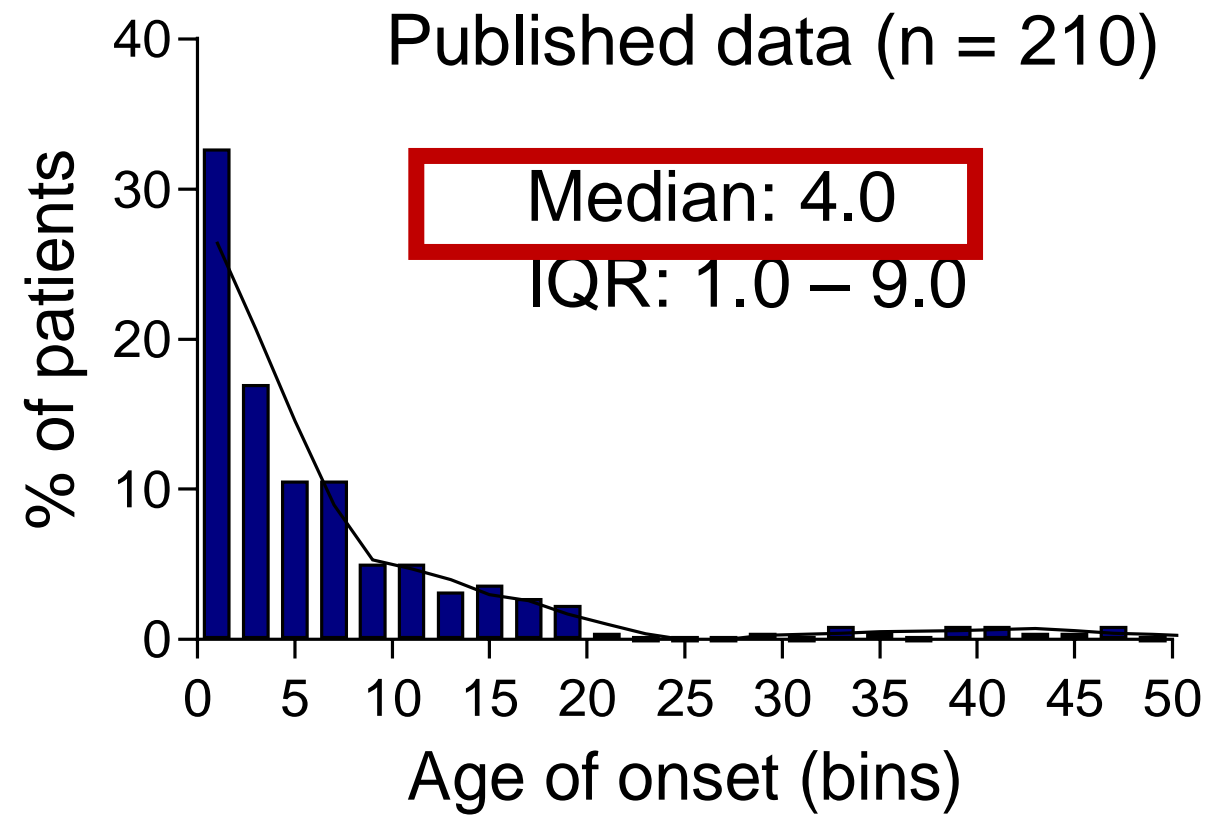
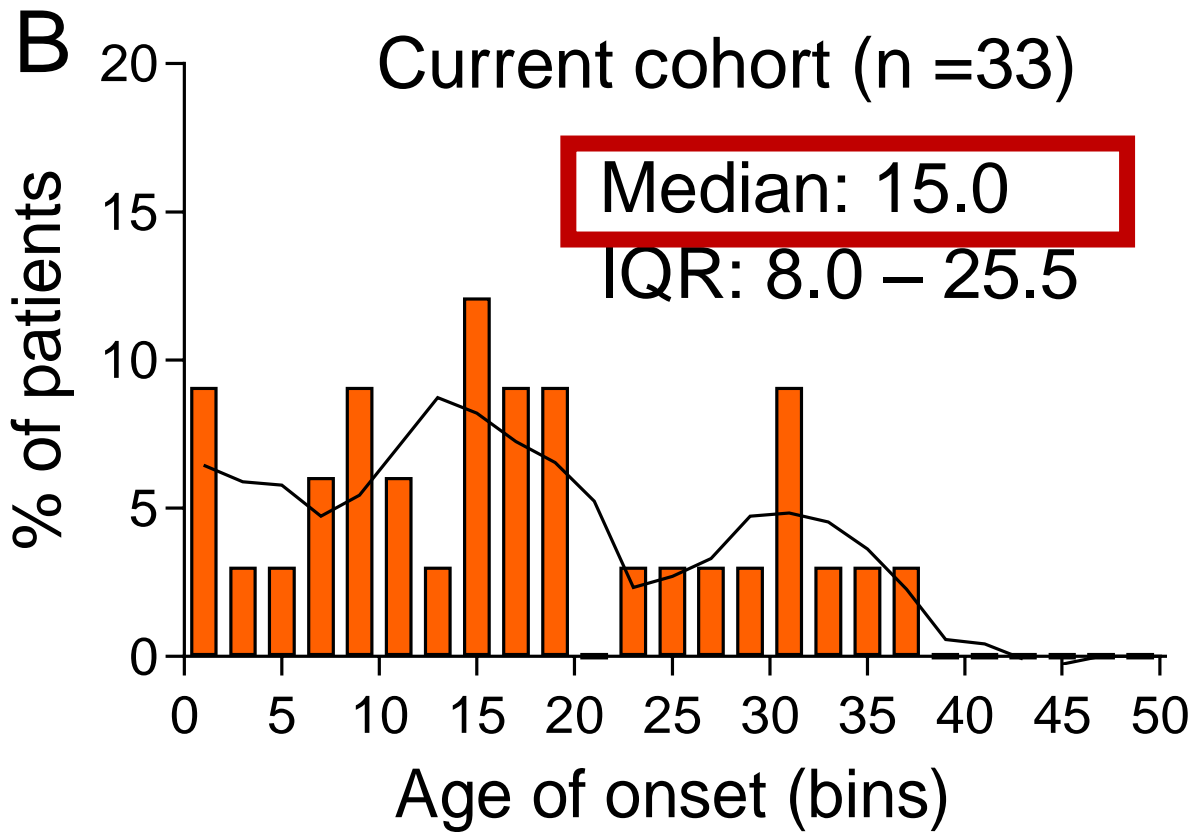




Deficiency of Adenosine Deaminase 2 in Adults and Children: Experience From India

Aman Sharma,¹  GSRSNK Naidu,¹ Vikas Sharma,¹ Saket Jha,² Aadhar Dhooria,³ Varun Dhir,¹ 
Prateek Bhatia,¹ Vishal Sharma,¹  Sagar Bhattad,⁴ K. G. Chengappa,⁵  Vikas Gupta,⁶
Durga Prasanna Misra,⁷  Pallavi Pimpale Chavan,⁸ Sourabh Malaviya,⁹ Rajkiran Dudam,¹⁰ Banwari Sharma,¹¹
Sathish Kumar,¹² Rajesh Bhojwani,¹³ Pankaj Gupta,¹ Vikas Agarwal,⁷ Kusum Sharma,¹ Manphool Singhal,¹
Manish Rathi,¹ Ritambhara Nada,¹ Ranjana W. Minz,¹ Ved Chaturvedi,¹⁴ Amita Aggarwal,⁷  Rohini Handa,¹⁵
Alice Grossi,¹⁶ Marco Gattorno,¹⁷  Zhengping Huang,¹⁸  Jun Wang,¹⁹ Ramesh Jois,²⁰ V. S. Negi,⁵
Raju Khubchandani,⁸ Sanjay Jain,¹ Juan I. Arostegui,²¹  Eugene P. Chambers,²² Michael S. Hershfield,²³
Ivona Aksentijevich,²⁴ Qing Zhou,¹⁹ and Pui Y. Lee²⁵



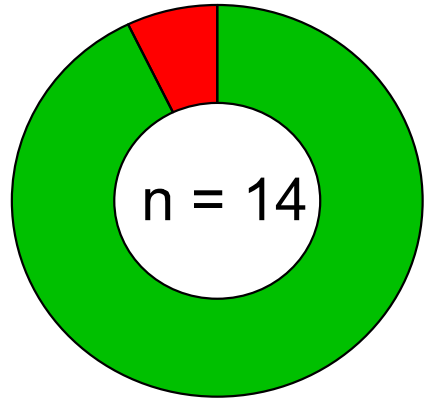


Sharma A et al. Arthritis Rheumatol. 2020 Sep 6.

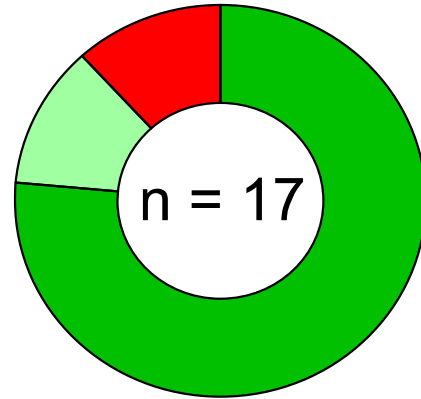


■ Resolved
 ■ Improved
 ■ Persistent
 ■ Relapse / New

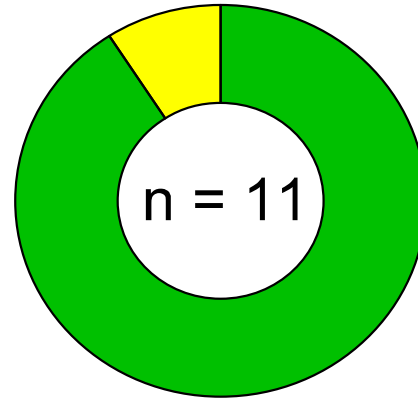
Recurrent fever



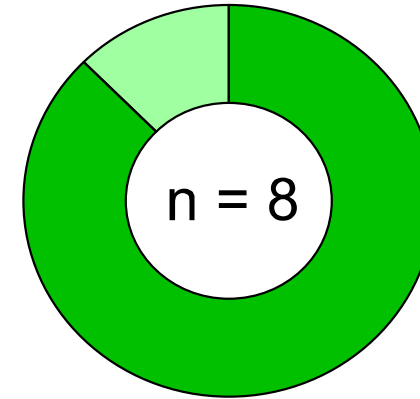
Skin



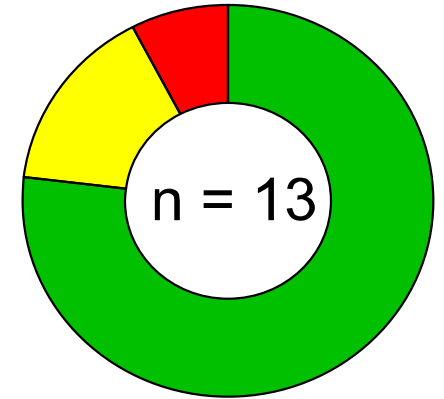
Gastrointestinal



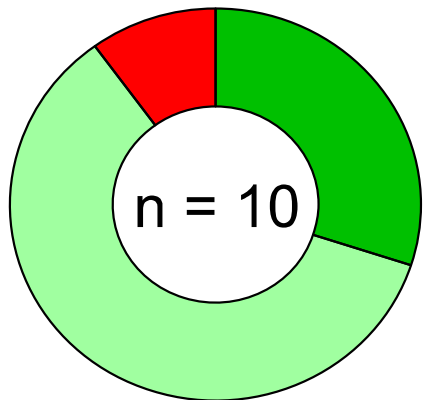
Musculoskeletal



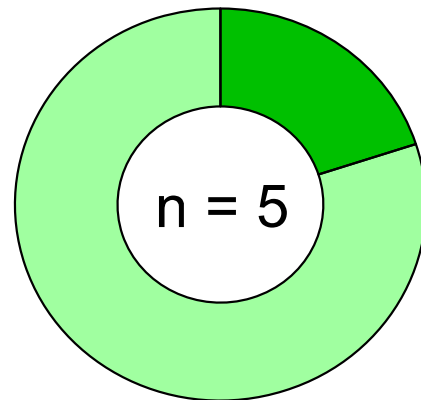
↑ ESR/CRP



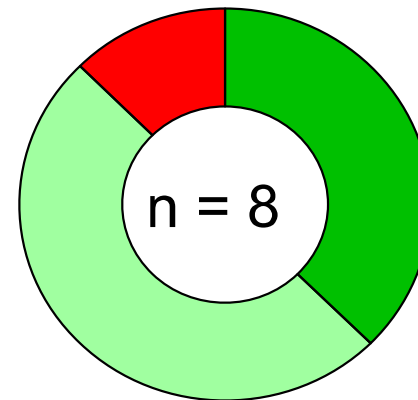
Ischemic stroke



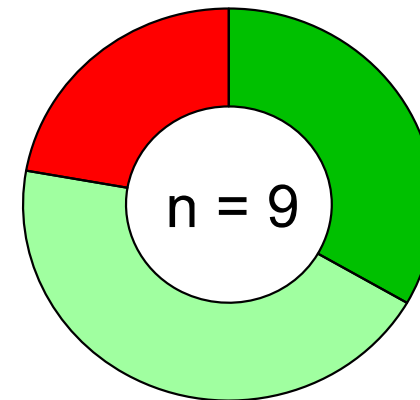
Hemorrhagic stroke



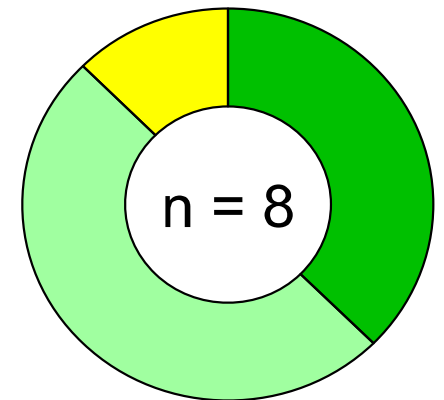
CNS neuropathy



PNS neuropathy



Anemia





Peter Grayson
@petercgrayson

2020

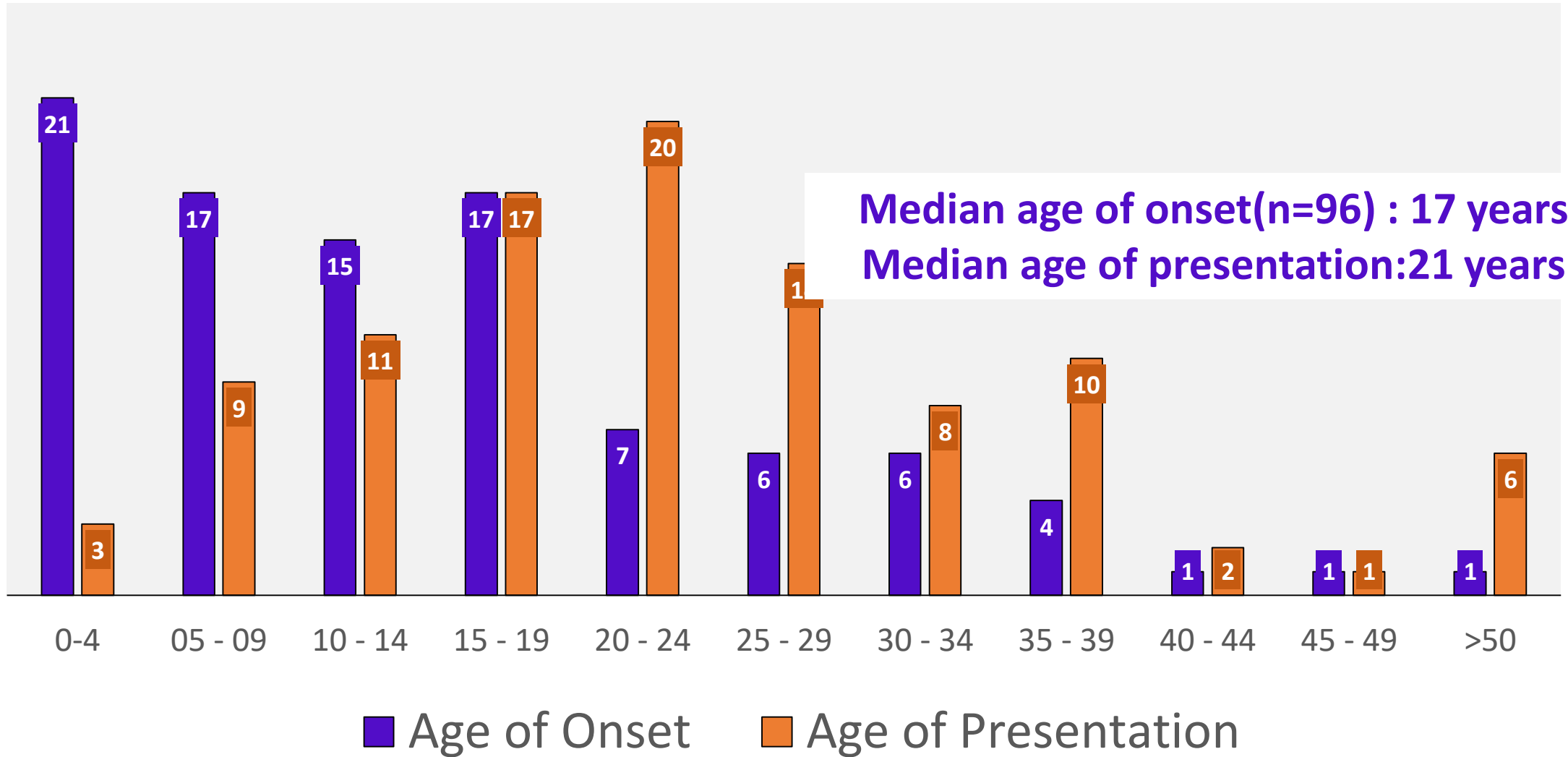
@Amansharmapgi giving the inaugural lecture about DADA2 in India at the 3rd International DADA2 conference. Great to see people around the world working together to treat rare diseases!
@dada2foundation #dada2conference



Peter C Grayson
Chief of the Vasculitis
Translational Research
Program, NIH



101 patients



Sharma A et al *Arthritis Rheumatol.* 2024; 76 (suppl 9).



Predictors of mortality

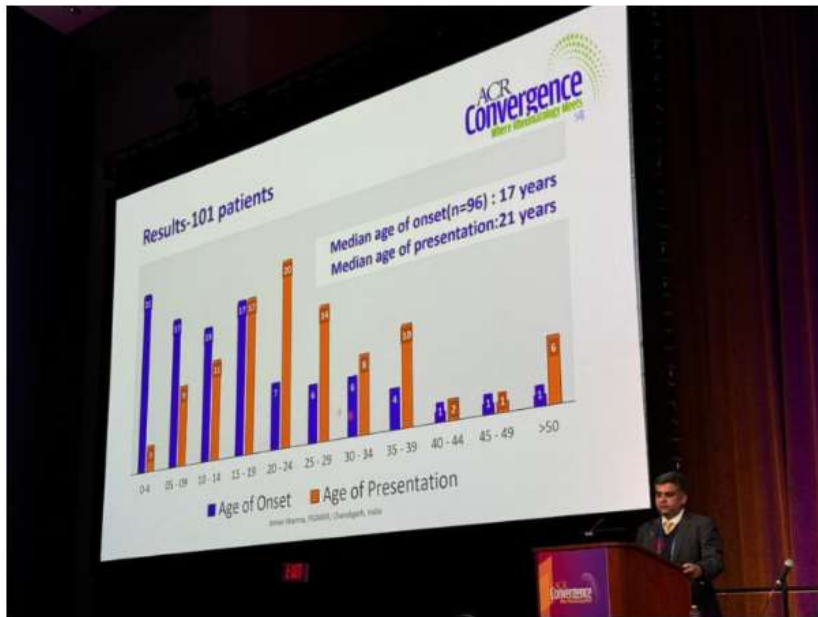
	Survivors- N (%)	Non- Survivors N (%)	p Value
Mesenteric Ischemia	9 (10.1)	5 (45.5)	0.007
Bowel Gangrene	6 (6.7)	2 (18.2)	0.213
CNS Involvement	47 (52.8)	5 (45.5)	0.352
Stroke	28 (31.5)	7 (63.6)	0.042
Cranial Neuropathy	23 (25.8)	4 (36.4)	0.349
PNS Involvement	36 (40.5)	4 (36.4)	1
Skin Involvement	52 (58.4)	8 (72.7)	0.518
Livedo Reticularis	31 (34.8)	6 (54.5)	0.284
Ocular Involvement	20 (22.4)	0 (0)	0.114
TNFi Use	61 (68.5)	4 (36.4)	0.047

Sharma A et al *Arthritis Rheumatol.* 2024; 76 (suppl 9).





Updates on the Indian Cohort



At the American College of Rheumatology (ACR) annual meeting last November, Dr. Aman Sharma, PGIMER, Chandigarh, India, presented the latest research out of the Indian cohort, perhaps **the largest cohort globally**. Over 40 physicians and researchers from around India participated in the study. The study, which analyzed data from 101 patients, underscores the importance of collaboration in advancing our understanding of this condition.





Consensus Statement | Pediatrics

Evaluation and Management of Deficiency of Adenosine Deaminase 2 An International Consensus Statement

Pui Y. Lee, MD, PhD; Brad A. Davidson, BS; Roshini S. Abraham, PhD; Blanche Alter, MD, MPH; Juan L. Arostegui, MD, PhD; Katherine Bell, MFA; Alexandre Belot, MD, PhD; Jenna R. E. Bergerson, MD, MPH; Timothy J. Bernard, MD, MScS; Paul A. Brogan, BSc, MBChB, MSc, PhD; Yackov Berkun, MD; Natalie T. Deutch, MS, CGC; Dimana Dimitrova, MD; Sophie A. Georgin-Lavialle, MD, PhD; Marco Gattorno, MD; Bodo Grimbacher, MD; Hasan Hashem, MD; Michael S. Hershfield, MD; Rebecca N. Ichord, MD; Kazushi Izawa, MD, PhD; Jennifer A. Kanakry, MD; Raju P. Khubchandani, MD; Femke C.C. Klouwer, MD, PhD; Evan A. Luton, MD; Ada W. Man, MD; Isabelle Meyts, MD, PhD; Joris M. Van Montfrans, MD, PhD; Seza Ozen, MD; Janna Saarela, MD, PhD; Gustavo C. Santos, MD; Aman Sharma, MD; Ariane Soldatos, MD, MPH; Rachel Sparks, MD; Troy R. Torgerson, MD, PhD; Ignacio Leandro Uriarte, MD; Taryn A. B. Youngstein, MB, BS, MD; Qing Zhou, PhD; Ivona Akseñtjievich, MD; Daniel L. Kastner, MD, PhD; Eugene P. Chambers, MD; Amanda K. Ombrello, MD; for the DADA2 Foundation



Chapel Hill 2012

- **Large Vessel Vasculitis (LVV)**

- Takayasu Arteritis (TAK)
- Giant Cell Arteritis (GCA)

- **Medium Vessel Vasculitis (MVV)**

- Polyarteritis Nodosa (PAN)
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- **Small Vessel Vasculitis (SVV)**

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- Granulomatosis with Polyangiitis (GPA)
- Eosinophilic Granulomatosis with Polyangiitis (EGPA)

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- Behçet Disease (BD)
- Cogan's Syndrome

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- Cutaneous Arteritis
- Primary Angiitis of the CNS (PACNS)
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- **Vasculitis Associated with Systemic Disease**

- Lupus vasculitis
- Rheumatoid vasculitis
- Sarcoid vasculitis

- **Vasculitis with Probable Etiology**

- HBV/HCV-associated vasculitis
- Drug-associated immune complex vasculitis
- Drug-associated ANCA-associated Vasculitis
- Cancer associated vasculitis



Granulomatous Vasculitis



Aman Sharma, MD, MAMS, FICP, FACR, FRCP(London)^{a,*},
Sunil Dogra, MD, MAMS, FRCP(London)^b, Kusum Sharma, MD, MAMS^c

Table 3
Comparison of different features of various vasculitides

Disease	ANCA	ENT	Lungs	Kidneys	Heart	Nerves	Skin
GPA	PR3 80%–95% MPO 5%–20% ANCA-Neg	+++	Nodules Infiltrates	++++	+	++	++
EGPA	MPO 40% PR3 35%	+	Asthma Infiltrates	++++	+	+++	+++
MPA	MPO 40%–80% PR3 35%	–	Infiltrates	+++	++	++++	+++
PAN	–	–	–	–	++	++++	+++
Cut PAN	–	–	–	–	–	+	++++

Abbreviations: ANCA-Neg, ANCA negative; Cut PAN, cutaneous polyarteritis nodosa; ENT, ear, nose, and throat.

Dermatol Clin 33 (2015) 475–487



Clinical features and long-term outcomes of 105 granulomatosis with polyangiitis patients: A single center experience from north India

Aman SHARMA,¹ Godasi S. R. S. N. K. NAIDU,¹ Manish RATHI,² Roshan VERMA,³ Manish MODI,⁴ Benzeeta PINTO,¹ Kusum SHARMA,⁵ Varun DHIR,¹ Manphool SINGHAL,⁶ Mahesh PRAKASH,⁶ Ritambhra NADA,⁷ Naresh K. PANDA³ and Ranjana W. MINZ⁸

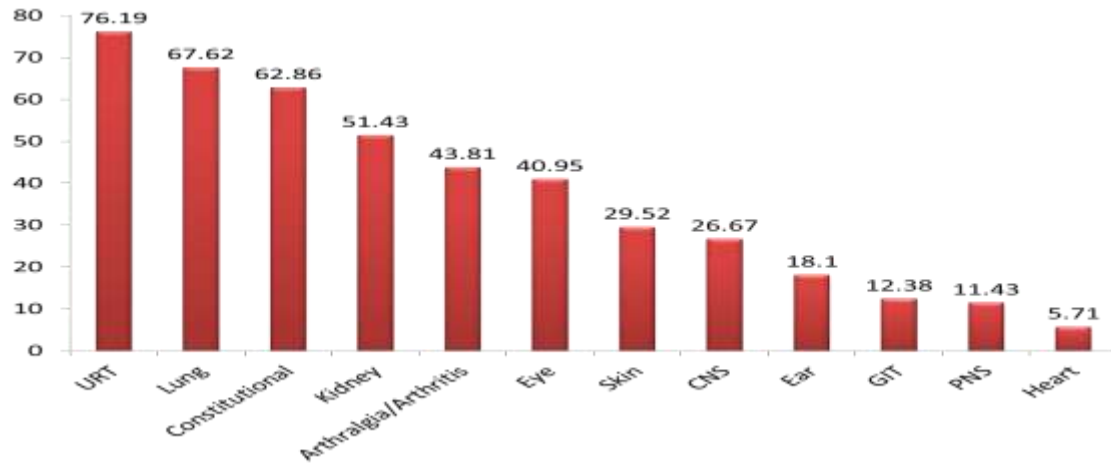


Table 1 Baseline characteristics of study population

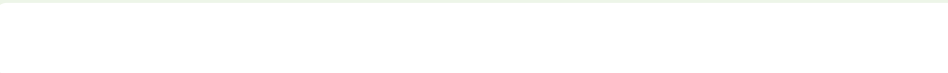

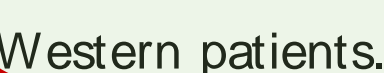
Number of patients	105
Males, <i>n</i> (%)	45 (42.86)
Females, <i>n</i> (%)	60 (57.14)
Mean age in years (range)	40.31 (15–70)
Mean duration of follow up in months (range)	37.21 (0–136)
ANCA positivity, <i>n</i> (%)	94 (89.52)
c-ANCA	86 (81.9)
p-ANCA	7 (8.57)
PR3	65 (61.9)
MPO	7 (6.67)
ANCA negativity, <i>n</i> (%)	11 (10.48)





Is granulomatosis with polyangiitis in Asia different from the West?

Godasi S. R. S. N. K. Naidu¹ | Durga Prasanna Misra²  | Manish Rathi³  | Aman Sharma¹ 

disease onset whereas Japanese patients are older. There are also . Japanese and Chinese patients are predominantly MPO-ANCA positive while Indian patients have predominant PR3-ANCA positivity. Renal involvement is less in India and Korea as compared to Japanese, Chinese and Western patients. .  lar between MPO-ANCA GPA and PR3-ANCA GPA.

Int J Rheum Dis. 2019;22:90–94.



Successful treatment outcomes in pregnant patients with ANCA-associated vasculitides: A systematic review of literature

Pawan Singh¹ | Aadhaar Dhooria² | Manish Rathi³  | Ritesh Agarwal¹ |
Kusum Sharma⁴ | Varun Dhir²  | Ritambhara Nada⁵ | Ranjana Minz⁶ | Vanita Suri⁷ |
Sanjay Jain² | Aman Sharma² 

Timing of diagnosis (n = 110)	n (%)
Before pregnancy	69 (62.7)
During pregnancy	32 (29.1)
After pregnancy	9 (8.2)

Duration of remission before conception in months, n = 35, median (range)	36 (3-120)
--	------------



Granulomatosis with Polyangiitis or post pulmonary tuberculosis: can CT chest help in differentiating in a tuberculosis endemic area?

Sharma A, Singhal M, Sen M, Rathi M, Dhir V, Sharma K, Minz RW, Singh S, Bambery P

	POST PRIMARY TUBERCULOSIS	GRANULOMATOSIS WITH POLYANGIITIS-
1. Nodules	Centrilobular nodules surrounded with branching linear structures	Widespread distribution, no lobar predilection
	Ill-defined margins	Well defined with clear margins
2. Consolidation and GGOs	Predilection for apical and posterior segment of upper lobe and superior segment of lower lobe	No lobar or zonal predilection
	Focal or patchy Heterogenous consolidations	Dense consolidations with ill defined margins
2. Cavities	Thick or thin walled	Thick or thin walled
	Central cavitation is seen	Cavitation- eccentric (12.5%) or centric (22.5%)



Prevalence of Antineutrophil Cytoplasmic Antibodies and Antinuclear Antibodies in Patients with Pulmonary Tuberculosis: A Tertiary Care Center Experience from North India



Yogesh Chander¹, Nupoor Acharya², GSRSNK Naidu³, Manish Rathi⁴, Ranjana Minz⁵, Sanjay Jain⁶, Digambar Behera⁷, Aman Sharma^{8*}

Antibodies tested	N=89
Anti-MPO	0
Anti-PR3	7 (7.8)
Anti-elastase	86 (96.6)
Anti-lactoferrin	0
ANA	6 (6.7)

ANA, antinuclear antibody; ATT, antitubercular therapy; MPO, myeloperoxidase; PR3, proteinase-3; PTB, pulmonary tuberculosis; ZN, Ziehl-Neelsen





Contents lists available at [ScienceDirect](https://www.sciencedirect.com)

Autoimmunity Reviews

journal homepage: www.elsevier.com/locate/autrev



Review

Indian Rheumatology Association guidelines for the management of ANCA associated vasculitis



Aadhaar Dhooria^{a,1}, G.S.R.S.N.K. Naidu^{b,1}, Durga Prasanna Misra^c, Benzeeta Pinto^d, M.B. Adarsh^e, Saket Jha^f, Rajiv Ranjan Kumar^g, Arghya Chattopadhyay^h, Vikas Sharmaⁱ, Debashish Mishra^j, Nupoor Acharya^k, Sakshi Mittal^l, Siddharth Jain^m, Joydeep Samantaⁿ, Chengappa Kavadiachanda^o, Sahajal Dhooria^p, Raja Ramachandran^q, Ramesh Jois^r, Banwari Sharma^s, Canchi Balakrishnan^t, Vineeta Shobha^d, Uma Kumar^u, Vikas Agarwal^c, B.G. Dharmanand^r, Rohini Handa^v, Aman Sharma^{b,*}



Summary of Recommendations

Condition	Agents for Remission Induction	Agents for Remission Maintenance
Active GPA / MPA*	Glucocorticoids+ Rituximab or IV CYC	Rituximab > Azathioprine
Severe EGPA	Glucocorticoids+ IV CYC or Rituximab	Low dose Glucocorticoids+ Azathioprine / Methotrexate / MMF
Non-severe EGPA	Glucocorticoids+ Mepolizumab >Azathioprine / Methotrexate / MMF	Low dose Glucocorticoids

***Special considerations for active GPA / MPA:** Prefer rituximab in relapsing disease; Consider PLEX for severe renal disease (serum Creatinine >5.8 mg/dl) in addition to other remission induction agents



Experience with similar biologic rituximab in 77 patients of granulomatosis with polyangiitis—a real-life experience


Sakshi Mittal¹ & G. S. R. S. N. K. Naidu¹ & Saket Jha¹ & Manish Rathi² & Ritambhara Nada³ & Ranjana W Minz⁴ & Kusum Sharma⁵ & Varun Dhir¹ & Sanjay Jain¹ & Aman Sharma¹ 

Table 4 Comparison of outcomes with use of rituximab for remission induction between AAV trials and our cohort

Name of the study	RAVE 5	RITUXVAS [4]	French vasculitis group [19]	Present study
Trial design	Multicentre, double blind RCT – rituximab versus oral CYC	Multicentre, open label RCT – rituximab versus i.v. CYC	Multicentre, retrospective	Singles centre, retrospective
Study population (rituximab arm)	99 AAV (76 GPA)	33 AAV (18 GPA)	80 AAV, 73 received rituximab induction. 70 (88%) GPA	60 GPA patients
Rituximab dosing schedule	375 mg/m ² weekly × 4	375 mg/m ² weekly × 4 + i.v. cyclophosphamide 15 mg/kg with 1st and 3rd infusion	375 mg/m ² weekly × 4, 1 g every 2 weeks for 1 month	1 g 2 doses at 0 and 15 days*
Rituximab indication	48 primary induction, 52 re-induction at relapse	33 primary induction, 0 re-induction	1 primary induction, 72 re-induction	25 primary induction, 35 re-induction at relapse
BVAS	BVAS/WG mean 8.5 ± 3.2	BVAS 2003 mean 19.65 ± 1.23 ^a	Median BVAS-v3 7 (IQR 5–12)	Median BVAS-v3 12 (IQR 6–21.5)
Renal	65 (66%)	33 (100%)	44 (55%)	22 (37%)
Pulmonary	51 (52%)	0	57 (71%)	40 (77%)
Remission at 6 months	63/99 (64%) over all 46/73 (63%) in GPA	25/33 (76%) sustained remission ^a	51/77 (66%)	36/60 (60%)
Relapses at 6 months	13	1		14
Grade 3 or more infection	7	7 ^a	7	15 ^c
Malignancy				
At 6 months	1	-	-	0
Beyond 6 months	5	2	-	0
Mortality at 6 months	1 (1%)	6 (18.2%) ^a	3 (3.8%)	7 (11.7%) ^b



Impact of renal involvement on survival in ANCA-associated vasculitis

Manish Rathi² · Benzeeta Pinto¹ · Aadhaar Dhooria¹ · Vinay Sagar¹ · Tarun Mittal² ·
Roopa Rajan³ · Varun Dhir¹ · Susheel Kumar¹ · Kusum Sharma⁴ · Ritambhra Nada⁵ ·
Surjit Singh¹ · Ranjana Walker Minz⁶ · Aman Sharma¹

	Our data	Booth et al. [17]	Hilhorst et al. [15]
No. of patients	51	80	246
M:F	1.04:1	1:1	1.3:1
Age	45	63	66
GPA	35 (68.6 %)	32 (40 %)	120 (49 %)
MPA	11 (21.6 %)	28 (35 %)	82 (33 %)
ANCA neg	6 (11.7 %)	6 (7.5 %)	8 %
Mean creatinine	3.6 mg/dl	4.35 mg/dl	3.87 mg/dl
Relapse	21.8 %	33 %	34 %
Mortality	17 (19.5 %)	21 (26 %)	18 %
ESRD	1 (1.96 %)	18 (23 %)	68 (28 %)

M male, *F* female, *GPA* granulomatosis with polyangiitis, *MPA* microscopic polyangiitis, *ESRD* end-stage renal disease



Pauci-immune glomerulonephritis: does negativity of anti-neutrophilic cytoplasmic antibodies matters?

Aman SHARMA,^{1,*} Ritambra NADA,² Godasi S. R. S. N. K. NAIDU,³ Ranjana W. MINZ,⁴

Table 5 Treatment outcomes at 6 months

N (%)	ANCA- positive	ANCA- negative	<i>P</i> - value
Improved	27 (62.8)	10 (37.1)	0.02
Deteriorated	6 (14.0)	11 (40.7)	0.006
Death	10 (23.2)	6 (22.2)	0.44
Renal replacement therapy (RRT) at presentation	23 (45.1)	19 (57.6)	0.14
Dialysis-independent	16 (69.6)	6 (31.6)	
Dialysis-dependent	7 (30.4)	13 (68.4)	0.009

N=84

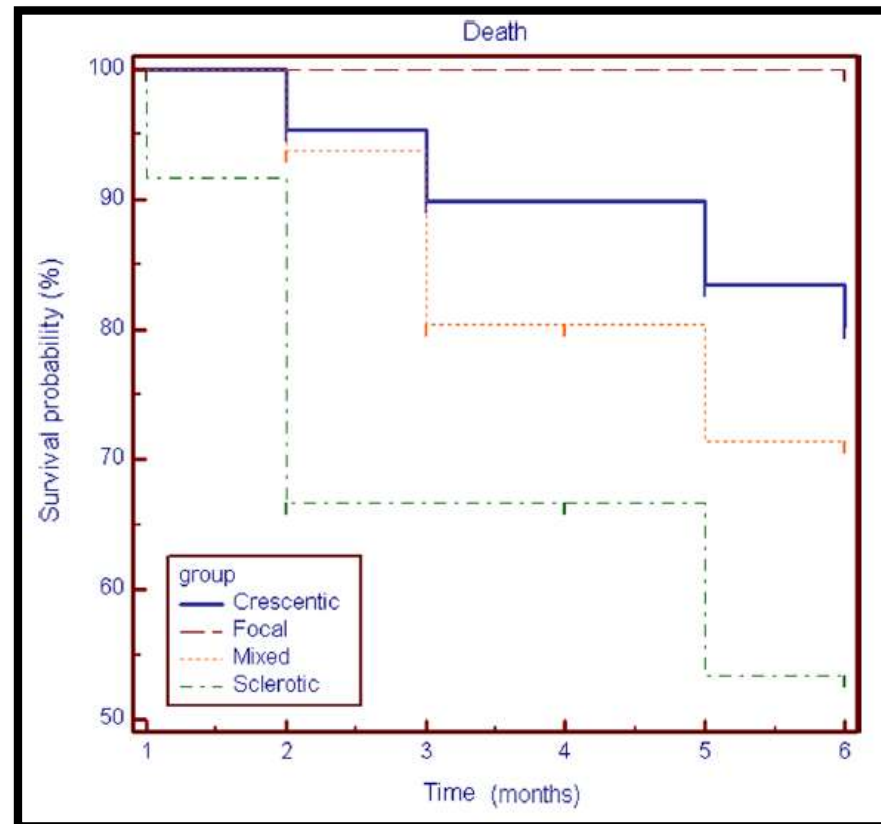
ANCA, anti-neutrophilic cytoplasmic antibodies.
Bold values indicate significant *P* values.

Sharma A et al. Int J Rheum Dis 2015



Histopathological classification of pauci-immune glomerulonephritis and its impact on outcome

Godasi S. R. S. N. K. Naidu · Aman Sharma ·
Ritambra Nada · Harbir Singh Kohli · Vivekanand Jha ·
Krishan Lal Gupta · Vinay Sakhuja · Manish Rathi



Rheumatol Int (2014) 34:1721–1727





Predominance of PR3 specific immune response and skewed TH17 vs. T-regulatory milieu in active granulomatosis with polyangiitis



Lekha Rani^a, Ranjana W. Minz^{a,*}, Aman Sharma^b, Shashi Anand^a, Dheeraj Gupta^c, N.K. Panda^d, V.K. Sakhuja^e

Increased ROR- γ t and decreased FOXP3 in active disease

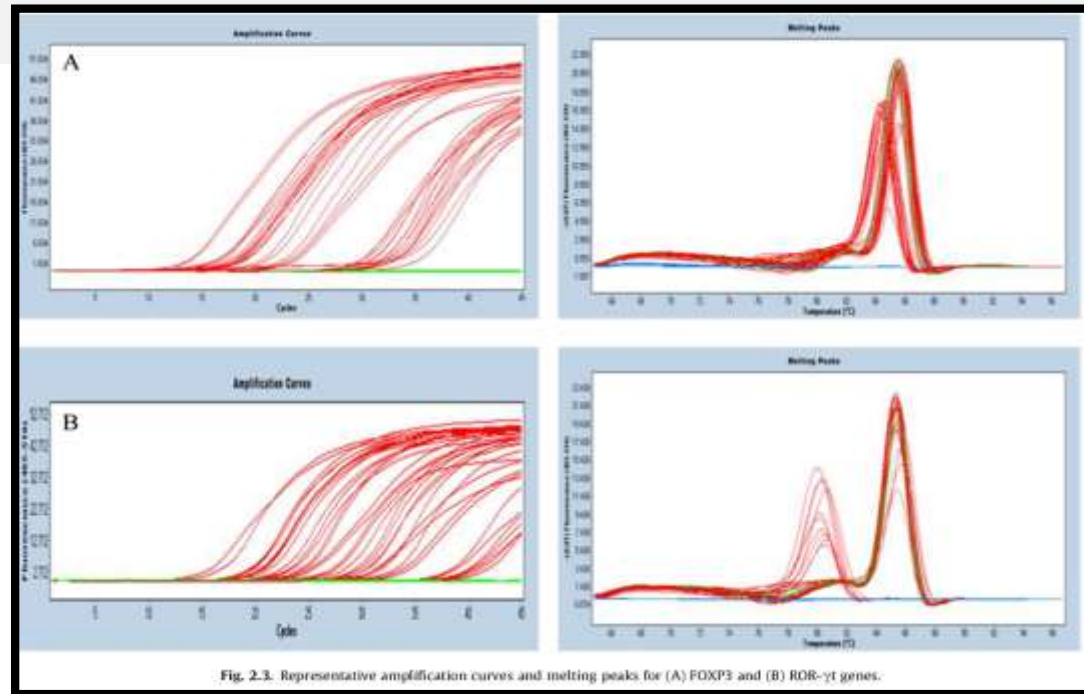


Fig. 2.3. Representative amplification curves and melting peaks for (A) FOXP3 and (B) ROR- γ t genes.

Cytokine. 2015 Feb;71(2):261-7.



Serum proteomic profiling in granumomatosis with polyangiitis using two-dimensional gel electrophoresis along with matrix assisted laser desorption ionization time of flight mass spectrometry

Lekha RANI,¹ Ranjana W. MINZ,¹ Amit ARORA,² Monica KANNAN,³ Aman SHARMA,⁴

Differentially expressed in active dss

- ✓ haptoglobin,
- ✓ Hp2, and
- ✓ NKp65,
- ✓ VDBP and
- ✓ Prolow-density lipoprotein receptor-related protein 1.



Distinct HLA and non-HLA associations in different subtypes of ANCA-associated vasculitides in North India

Jagdeep Singh¹ | Aman Sharma²  | Lekha Rani¹ | Navchetan Kaur¹ | Shashi Anand¹ |
Biman Saikia¹ | Saket Jha² | Ritambhra Nada³ | Ranjana Walker Minz¹ 

Int J Rheum Dis. 2020

Increased gene expression of B cell-activating factor of tumor necrosis factor family, in remitting antineutrophil cytoplasmic antibody-associated vasculitis patients

Saket Jha¹  | Jagdeep Singh²  | Ranjana W. Minz²  | Aadhar Dhooria¹  |
GSRSNK Naidu¹  | Rajiv Ranjan Kumar¹  | Manish Rathi³ | Sanjay Jain¹ |
Shashi Anand² | Aman Sharma¹ 

Int J Rheum Dis. 2022



Chapel Hill 2012

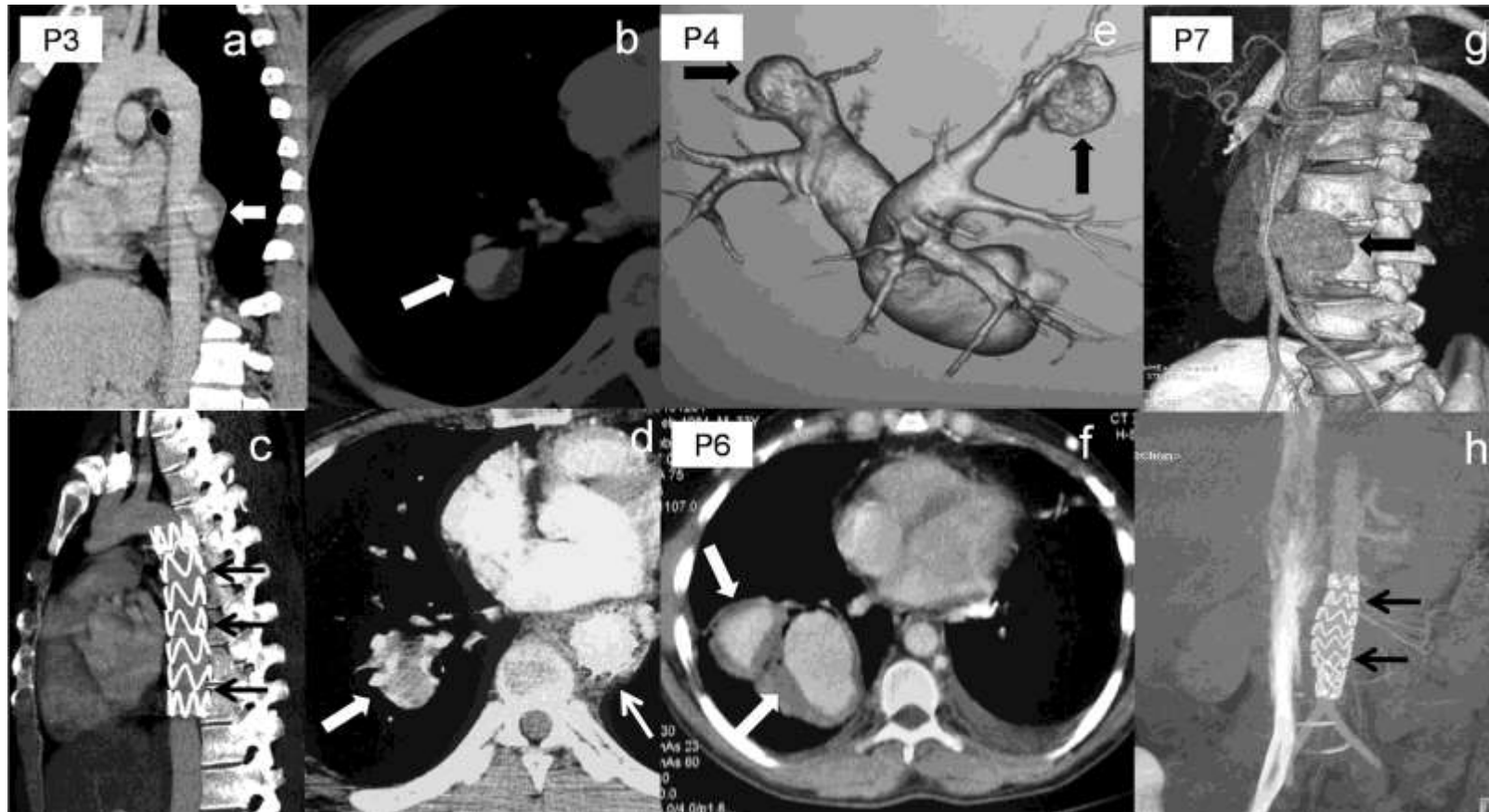
- **Large Vessel Vasculitis (LVV)**
 - Takayasu Arteritis (TAK)
 - Giant Cell Arteritis (GCA)
- **Medium Vessel Vasculitis (MVV)**
 - Polyarteritis Nodosa (PAN)
 - Kawasaki Disease (KD)
- **Small Vessel Vasculitis (SVV)**
 - **ANCA Associated Vasculitis**
 - Microscopic Polyangiitis (MPA)
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 - Eosinophilic Granulomatosis with Polyangiitis (EGPA)
 - **Immune complex SVV**
- **Variable Vessel Vasculitis (VVV)**
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 - Cogan's Syndrome
- **Single Organ Vasculitis (SOV)**
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- **Vasculitis Associated with Systemic Disease**
 - Lupus vasculitis
 - Rheumatoid vasculitis
 - Sarcoid vasculitis
- **Vasculitis with Probable Etiology**
 - HBV/HCV-associated vasculitis
 - Drug-associated immune complex vasculitis
 - Drug-associated ANCA-associated Vasculitis
 - Cancer associated vasculitis

Arthritis Rheum 2013; 65: 1-11



Clinical presentation and treatment outcomes of arterial involvement in Behçet's disease: a single-centre experience

Arghya Chattopadhyay¹  · Siddharth Jain¹  · G. S. R. S. N. K. Naidu¹  · Varun Dhir¹  · Bharath Chhabria²  · Nupoor Acharya¹  · Valliappan Muthu²  · Manphool Singhal³  · Ajay Savlania⁴  · Rajesh Vijayvergiya⁵  · Sanjay Jain¹  · Aman Sharma¹ 



Rheumatology International 202



Chapel Hill 2012

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Arthritis Rheum 2013; 65: 1-11







Connective Tissue Disorder-Associated Vasculitis

Aman Sharma¹ & Aadhaar Dhooria¹ & Ashish Aggarwal¹ &
Manish Rathi² & Vinod Chandran^{3,4,5}

Connective tissue disease	Incidence/prevalence ^a	Organ involvement
Rheumatoid vasculitis	3.6/million (population)	Eye, skin, lung, PNS, CNS, GI, kidney
Lupus vasculitis	11–36 %	Eye, skin, lung, PNS, CNS, GI, kidney
Sarcoid vasculitis	N.A.	Lung, skin, medium and large vessels
Systemic sclerosis-associated vasculitis	N.A.	Renal (with AAV)
Sjogren's syndrome-associated vasculitis	5–32 %	Skin, PNS
Relapsing polychondritis-associated vasculitis	N.A.	Skin, eye, aorta, coronary vessels, CNS
Idiopathic inflammatory myositis	9.2–30.4 %	Skin, GI



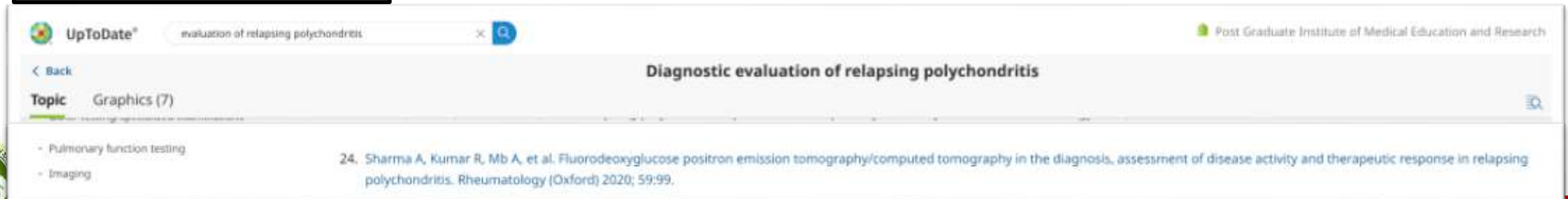
Fluorodeoxyglucose positron emission tomography/ computed tomography in the diagnosis, assessment of disease activity and therapeutic response in relapsing polychondritis

Aman Sharma ^{1,*}, Rajender Kumar^{2,*}, Adarsh MB ¹, GSRSNK Naidu ¹,
Vikas Sharma¹, Apurva Sood², Varun Dhir¹, Roshan Verma³,
Harmandeep Singh², Anish Bhattacharya², Sanjay Jain ¹ and
Bhagwant Rai Mittal²

- Diagnosing **‘hidden’ disease** in airways and large blood vessels
- Guiding management decisions
- Fair correlation with clinical symptoms and FDG uptake

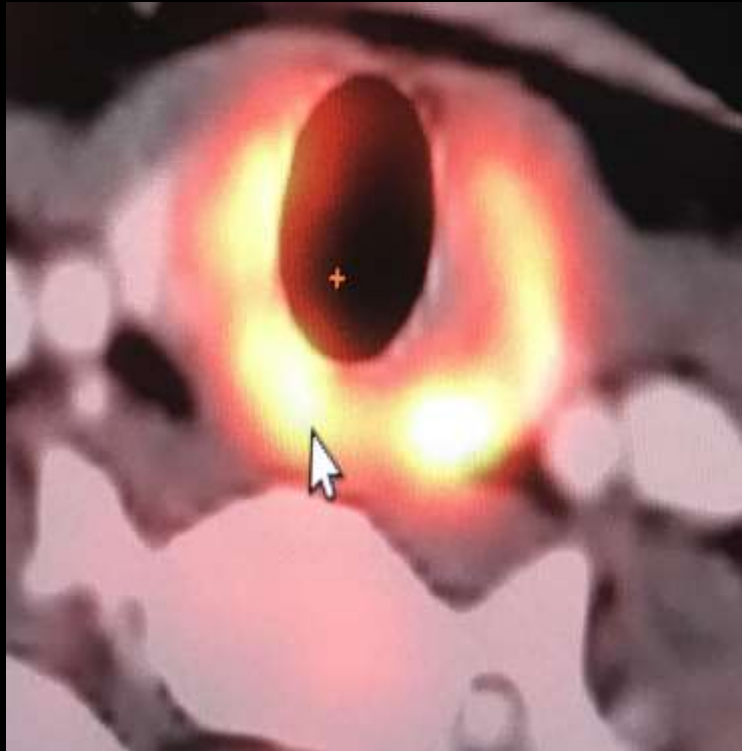
UpToDate.Com

Hochberg’s Textbook of Rheumatology

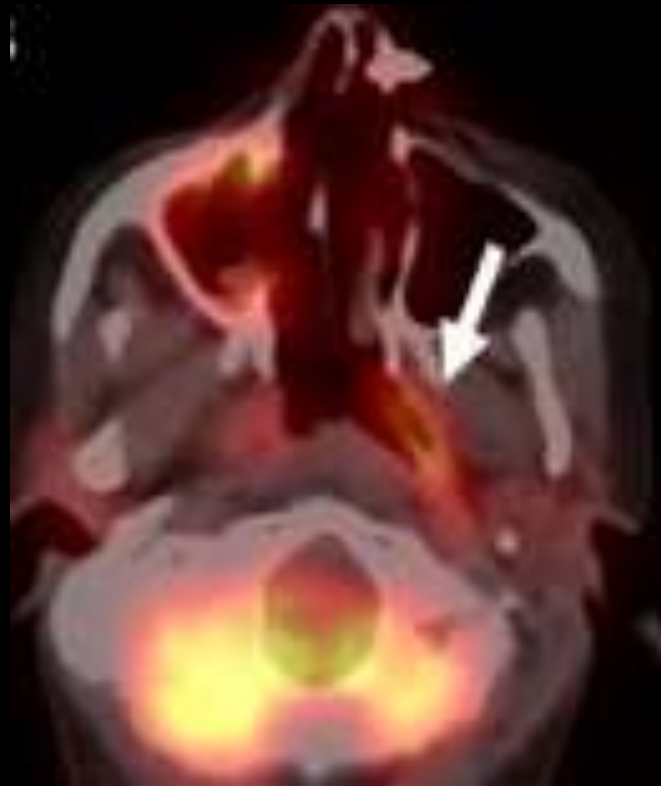


The screenshot shows a search result on UpToDate for 'evaluation of relapsing polychondritis'. The search bar contains the text 'evaluation of relapsing polychondritis'. The results page shows a list of topics, including 'Pulmonary function testing' and 'Imaging'. A specific reference is listed: '24. Sharma A, Kumar R, Mb A, et al. Fluorodeoxyglucose positron emission tomography/computed tomography in the diagnosis, assessment of disease activity and therapeutic response in relapsing polychondritis. Rheumatology (Oxford) 2020; 59:99.'

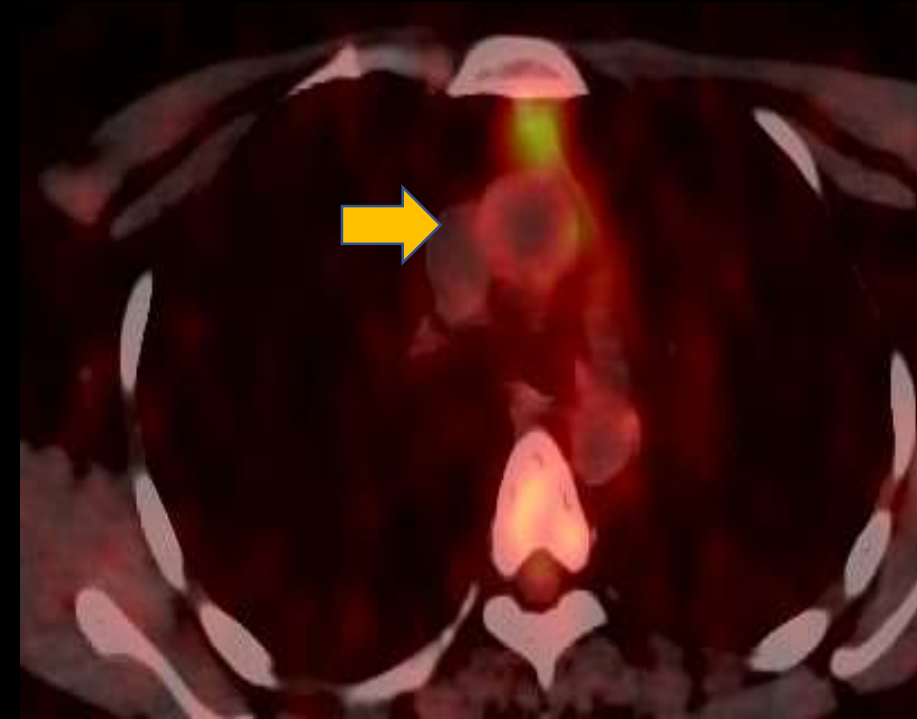
Extent of the disease



Airways



Eustachian tube



Aorta



Disease Activity Parameters: RPDAI & RPDAM

The Relapsing Polychondritis Disease Activity Index: Development of a disease activity score for relapsing polychondritis

Laurent Arnaud ^{a,b,*}, Hervé Devilliers ^{a,c,1}, Stanford L. Peng ^d, Alexis Mathian ^{a,b,e},
Nathalie Costedoat-Chalumeau ^{a,e}, Jane Buckner ^d, Lorenzo Dagna ^f, Clement Michet ^g, Aman Sharma ^h,

Autoimmun Rev. 2012

Original article

The relapsing polychondritis damage index (RPDAM): Development of a disease-specific damage score for relapsing polychondritis

Philippe Mertz ^{a,b}, Alexandre Belot ^c, Ricard Cervera ^d, Tyng Yu Chuah ^e, Lorenzo Dagna ^f,
Laura Damian ^g, Debashish Danda ^h, David D'cruz ⁱ, Gerard Espinosa ^d, Camille Frances ^j,
David Jayne ^k, Kong Kok Ooi ^{l,m}, Eugene J. Kucharz ⁿ, Robert Lebovics ^o, Isabelle Marie ^p,
Guillaume Moulis ^{q,r}, Stanford Peng ^s, Aman Sharma ^t, Noboru Suzuki ^u, Toshio Tanaka ^v,
Ronald Van Vollenhoven ^w, Jean Sibilia ^{a,b}, Jacques Eric Gottenberg ^{a,b}, François Chasset ^j,
Laurent Arnaud ^{a,b,*}

Joint Bone Spine. 2019

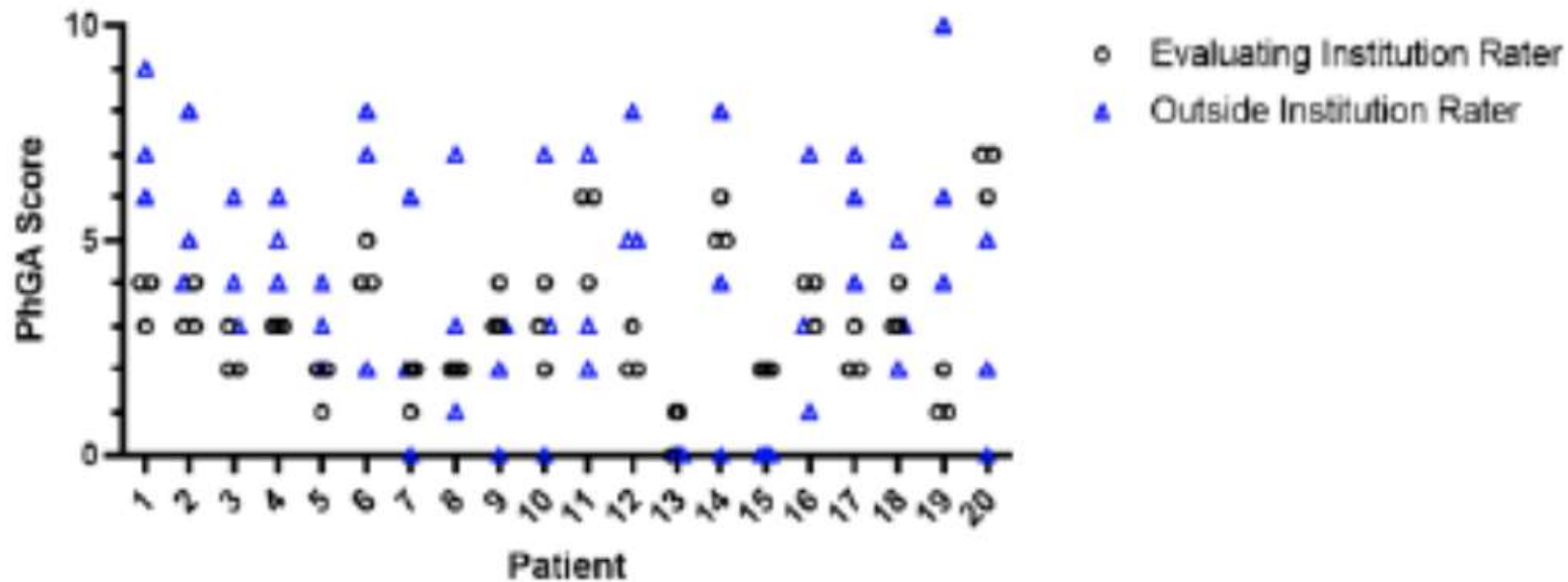


Physician Global Assessment as a Disease Activity Measure for Relapsing Polychondritis

Running Head: Physician Global Assessment for RP

Authors:

Emily Rose, BS¹, Marcela A. Ferrada, MD¹, Kaitlin A. Quinn, MD¹, Wendy Goodspeed, RN¹,
Laurent Arnaud, MD, PhD², Aman Sharma, MD³, Hajime Yoshifuji, MD, PhD⁴, Jeff Kim, MD⁵, Clint
Allen, MD⁶, Arlene Sirajuddin, MD⁷, Marcus Chen, MD⁷, Peter C. Grayson, MD, MSc¹

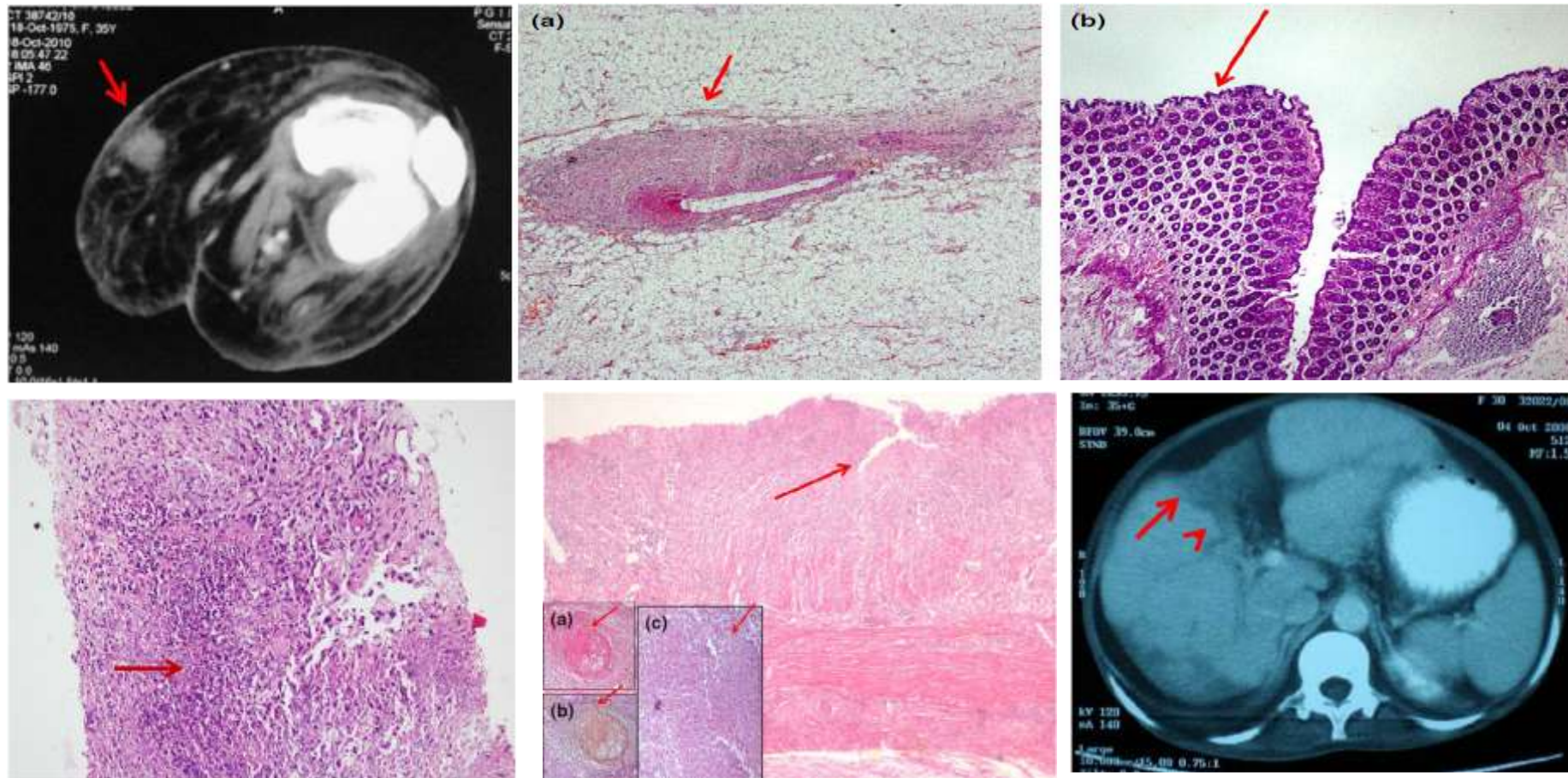


Arthritis Care Res (Hoboken). 2021 Feb 5;10.1002/acr.24



Uncommon presentations of primary systemic necrotizing vasculitides: the Great Masquerades

Aman SHARMA,¹ Dharmesh GOPALAKRISHAN,¹ Ritambhra NADA,² Susheel KUMAR,¹



Conclusions:

Our group has described

- **Clinical spectrum, validation of criteria, lab manifestations and imaging details of various vasculitis disorders**
- **DADA2 occurs in adults**
- **Expanded spectrum of DADA2 and VEXAS syndrome**
- **Genetic cytokine and proteomics studies in AAV**
- **Data driven Indian Guidelines**



Acknowledgements

Pradeep Bambery

GSRSNK Naidu

Ritambhra Nada

Ranjana W Minz

Rajender Kumar

Raja Ramchandran

Rajesh Vijayvergiya

Sanjay Jain

Manphool Singhal

Kusum Sharma

Vishal Sharma

Durga Prasanna Misra

Vikas Agarwal

Joydeep Samanta

Saket Jha

Arghya Chattopadhyay

Aaadhar Dhooria

Prateek Deo

Chirag R Kopp

Siddharth Jain

Vikas Sharma

Benzeeta Pinto

Lekha Rani

Jagdeep Singh

Tarun Mittal

Roopa Rajan

Pui Lee

Chip Chambers

Qing Zhou

Juan I Arostegui

Michael S. Hershfield

Ivona Aksentijevich

Marcella Ferrada

Peter Grayson

David Beck



Thank you



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