

Characteristics of an Internet-Based, International Cohort of Patients with a Self-Reported Diagnosis of Urticarial Vasculitis

Jason M Springer¹, Tanaz A Kermani², Dianne Shaw³, Kalen Larson⁴, Cristina Burroughs⁵, and Peter A. Merkel⁶,
for the Vasculitis Patient-Powered Research Network

¹Vanderbilt University Medical Center, Nashville, TN; ²University of California Los Angeles, Los Angeles, CA; ³Vasculitis Foundation, North Carolina; ⁴Vasculitis Foundation, Kansas City, MO; ⁵University of South Florida, Tampa, FL; ⁶University of Pennsylvania, Philadelphia, PA

INTRODUCTION

Urticarial vasculitis (UV) is a rare disease (annual incidence of < 1 per million). This rarity makes it difficult to conduct large trials.

AIMS

To describe and validate an international cohort of patients with UV based on patient-reported data.

METHODS

- Internet-based longitudinal registry
- Enrollment period: 11/2014-2/2022
- All patients with a self-reported diagnosis of urticarial vasculitis were included
- Standardized online data collection forms
- 2012 Chapel Hill Consensus Conference (CHCC) definition of hypocomplementemic urticarial vasculitis (HUVS) met if all three of the following were present:
 - a) Biopsy showing vasculitis
 - b) Low serum levels of C3 or C4
 - c) Any of the following manifestations: kidney, joint, obstructive lung disease, or

RESULTS

Baseline Demographics		Means of Diagnosis	
Enrollment	102		N (%)
Female (%)	92 (90)	Biopsy results	84 (82)
Mean age at symptom onset (SD)	40.5 (15.0)	Based on symptoms	58 (57)
Mean age at diagnosis (SD)	44.1 (13.2)	Laboratory testing of the blood	45 (44)
Low C3 or C4 (%)	31/59 (53)	Radiographic testing	7 (7)
Skin biopsy done (%)	89/99 (90)	Other	4 (4)
Skin biopsy with vasculitis (%)	77/89 (87)	Unsure	2 (2)
Met CHCC criteria for HUVS (%)	23/31 (74)		
Symptoms/Manifestations		Medication Use (Ever)	
	N (%)		N (%)
Hive-like rash	102/102 (100)	Glucocorticoids	89 (87)
Severe joint pain or swelling	77/93 (83)	Dapsone	27 (26)
Severe muscle pain	73/94 (78)	Hydroxychloroquine	26 (25)
Nerve damage	56/77 (73)	Colchicine	23 (23)
Abdominal pain	49/79 (62)	Methotrexate	19 (19)
Fever	32/78 (41)	Antihistamines	18 (18)
Inflammation in one or both eyes	32/87 (37)	Azathioprine	16 (16)
Lung involvement	28/84 (33)	Omalizumab	8 (8)
COPD or Asthma	37/91 (41)	Aspirin	8 (8)
Weight loss	19/92 (21)	Mycophenolate mofetil	7 (7)
Kidney involvement	17/88 (19)	Cyclosporine	5 (5)
CNS involvement	6/80 (8)	Rituximab	4 (4)
Pericarditis	5/74 (7)	TNF inhibitors	3 (3)
Thrombosis	5/87 (6)	Plasmapheresis	1 (1)
Any systemic manifestation	85/102 (83)		
Positive ANA	33/64 (52)	Physician Managing Vasculitis (Top 4)	
			n (%)
		Rheumatologist	52 (51)
		Primary Care Physician	35 (34)
		Dermatologist	26 (25)
		Allergist/Immunologist	16 (16)

DISCUSSION

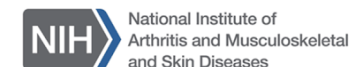
- The diagnosis of urticarial vasculitis is clinicopathological, based on presence of urticaria and histologic confirmation
- All patients in this cohort reported urticarial lesions, the majority of which were confirmed by biopsy
- All clinical manifestations reported are consistent with physician-reported cohorts

CONCLUSIONS

Internet-based cohorts incorporating patient-reported data are a means for future conduct of clinical trials in extremely rare diseases, such as urticarial

ACKNOWLEDGEMENTS

Supported by the



The VPPRN is supported in part by the Vasculitis Foundation