The Impact of Neurologic Involvement in ANCA-Associated Vasculitis on Self-Reported Health-Related Quality of Life: Data from the Vasculitis Patient-Powered Research Network

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BACKGROUND

- Neurologic involvement (NI) is common in ANCA-Associated Vasculitis (AAV), ranging from 22 to 54% in GPA, 34 to 72% in MPA, and up to 75% in EGPA
- NI in patients with AAV has a negative impact on multiple domains of illness and health-related quality of life (HRQoL)
- Most data on NI in AAV are derived from clinician-reported data
- This project assessed the prevalence and impact of NI on HRQoL in patients with AAV enrolled in the Vasculitis Patient-Powered Research Network (VPPRN), an online registry in which patients provide data

METHODS

- Patients with GPA, MPA, or EGPA enrolled in the VPPRN
- Data captured from 2014-2022 from patients at least 18 years old at enrollment
- Patient subsets were compared according to presence or absence of NI
- HRQoL assessments included several PROMIS domains:
  - Fatigue, Depression, Sleep Disturbance , Pain Interference, Physical Function, Social Isolation, and Anxiety
  - PROMIS measures generate T-scores with the mean T-score = 50 with a standard deviation of 10 in the reference population (U.S. general population)

RESULTS

Table 1. Demographics of patients with or without NI

Variable		All Patients (n=1465)	With Neurologic Involvement (n=901)	Without Neurologic Involvement (n=564)	P-value	N
Age at Diagnosis (years)		42.0 [28.0; 54.0]	45.0 [32.0; 56.0]	40.0 [22.0; 53.0]	0.032	211
Gender	Female	1025 (70.1%)	623 (69.3%)	402 (71.4%)	0.426	1462
	Male	437 (29.9%)	276 (30.7%)	161 (28.6%)		
Race	Asian	28 (1.91%)	14 (1.55%)	14 (2.48%)		
	Black	19 (1.30%)	12 (1.33%)	7 (1.24%)		
	White	1335 (91.1%)	822 (91.2%)	513 (91.0%)		
	Other	83 (5.67%)	53 (5.88%)	30 (5.32%)		
Type of Vasculitis	EGPA	266 (18.2%)	121 (13.4%)	145 (25.7%)	<0.001	1465
	GPA	985 (67.2%)	634 (70.4%)	351 (62.2%)		
	MPA	214 (14.6%)	146 (16.2%)	68 (12.1%)		
Disease Duration (years)		8.63 [4.88;13.8]	8.59 [4.72;13.3]	8.72 [5.21;15.1]	0.162	1213
ANCA-Positive		1123 (86.7%)	703 (88.7%)	420 (83.5%)	0.010	1296

Continuous variables presented as Mean (SD) or Median [P25, P75], categorical variables presented as N (column %).

Figure 1. Patients with NI had higher scores for fatigue, depression, sleep disturbance, pain interference, and lower scores for anxiety, social, physical function

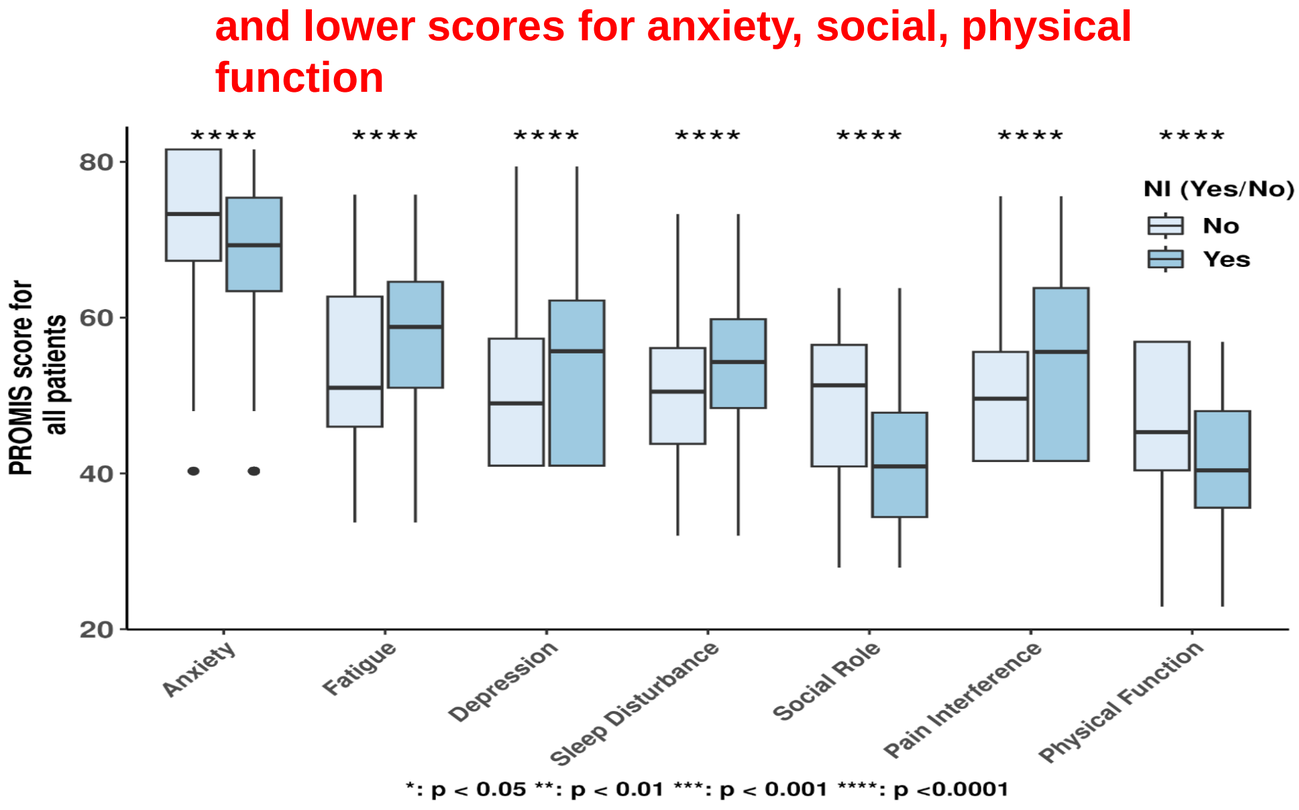

Table 2. Clinical characteristics of patients with or without NI

Variable	All Patients (n=1465)	With Neurologic Involvement (n=901)	Without Neurologic Involvement (n=564)	P-value	N
Constitutional Symptoms	994 (75.2%)	660 (81.5%)	334 (65.2%)	<0.001	1322
Musculoskeletal	1174 (83.0%)	815 (92.4%)	359 (67.5%)	<0.001	1414
Skin	838 (60.0%)	578 (67.5%)	260 (48.1%)	<0.001	1396
Mucous Membranes	515 (38.0%)	367 (44.9%)	148 (27.6%)	<0.001	1355
Eye	624 (45.4%)	401 (48.1%)	223 (41.2%)	0.015	1375
Ear, Nose, and Throat	1138 (83.1%)	751 (88.7%)	387 (74.1%)	<0.001	1369
Cardiovascular	123 (9.91%)	98 (13.7%)	25 (4.74%)	<0.001	1241
Gastrointestinal	37 (2.72%)	29 (3.55%)	8 (1.47%)	0.033	1362
Pulmonary	928 (66.0%)	616 (72.0%)	312 (56.8%)	<0.001	1405
Renal	905 (63.3%)	559 (63.8%)	346 (62.5%)	0.644	1430
Venous Thrombosis	187 (13.4%)	132 (15.7%)	55 (10.0%)	0.003	1393
Severity of Vasculitis	4.00 [1.00;5.00]	5.00 [2.00;6.00]	3.00 [0.00;5.00]	<0.001	1462

Continuous variables presented as Mean (SD) or Median [P25, P75], categorical variables presented as N (column %).

Table 3. Prevalence of NI in patients with AAV enrolled in VPPRN is higher compared to major cohorts

	VPPRN N=1465	DCVAS* N= 955	VCRC N= 1368	P value
Patients with AAV	62%	28%	34%	<0.001
Patients with GPA	64%	19%	25%	<0.001
Patients with MPA	68%	23%	22%	<0.001
Patients with EGPA	46%	65%	67%	<0.001

*DCVAS included patients with vasculitic neuropathy only

CONCLUSIONS

- NI in AAV is common and is associated with significant reductions in HRQoL
- The prevalence of NI as reported by patients compared to physician-reported data is higher for GPA and MPA, but lower for EGPA
- Compared to patients without NI, patients with NI
  - Are older at the time of diagnosis of AAV
  - Are more often positive for ANCA
  - Have more different manifestations of AAV
  - Report greater severity of their vasculitis
- The rate of renal involvement is the same among patients with or without NI
- Patients with AAV report reduced HRQoL in almost all domains compared to a reference general population

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