

SUPPLEMENTAL TABLES AND FIGURES

Supplemental Table 1: SLE-related Autoantibodies performed	
Autoantibody	Assay and Cut-offs
Anti-dsDNA	Anti-dsDNA titers and positivity were detected by a chemiluminescence immunoassay (CLIA) (Werfen, San Diego, USA) using a cut-off of >27 chemiluminescence units (CU). Positive was >35 (CU), while 27-35 (CU) was considered indeterminate (borderline).
Other SLE-specific autoantibodies	Using a Luminex 200 flow luminometer (Luminex, Austin, USA), ALBIA (FIDIS Connective13: TheraDiag, Paris, France) was performed to analyze SLE-related analytes, which included ribosomal P, Ro52/Tripartite Motif Protein 21 (TRIM21), SSA/Ro60, SSB/La, Sm, U1-RNP, and histones. A cut-off of >40 median fluorescence units (MFU) was considered positive.
Anti-phospholipid antibodies	Anti-phospholipid antibodies (IgG and IgM anticardiolipin and anti- β 2-glycoprotein-1) were measured using ELISA (Werfen, San Diego, USA). As per the revised Sapporo antiphospholipid syndrome classification criteria (1), a cut-off of >40 units for IgG/IgM anticardiolipin was considered medium to high positive while a cut-off of >20 units (>99 th percentile) was positive for IgG/IgM anti- β 2-glycoprotein-1 (1). All autoantibodies were measured at MitogenDx except for lupus anticoagulant, which was measured at Oklahoma Medical Foundation (Oklahoma City, OK) as previously described (2).

1. Miyakis S, Lockshin MD, Atsumi T, Branch DW, Brey RL, Cervera R, et al. International consensus statement on an update of the classification criteria for definite antiphospholipid syndrome (APS). *J Thromb Haemost*. 2006;4(2):295-306.
2. Hanly J, Urowitz M, Siannis F, Farewell V, Gordon C, Bae S, et al. Autoantibodies and neuropsychiatric events at the time of systemic lupus erythematosus diagnosis: results from an international inception cohort study. *Arthritis & Rheumatism: Official Journal of the American College of Rheumatology*. 2008;58(3):843-53.

Supplemental Table 2: Distribution of anti-KIF20B positivity at baseline between patients with and without NPSLE manifestations^a within 5 years of follow-up using Attribution Model B				
NPSLE Manifestations, %	Cohort (n=795)	KIF20B+ (n=237)	KIF20B- (n=558)	p-value
Headache	0.0%	0.0%	0.0%	-
Mood Disorders	6.4%	3.8%	7.5%	0.050
Seizures and/or Seizure Disorder	3.4%	3.4%	3.4%	0.983
Cognitive Dysfunction	3.0%	3.4%	2.9%	0.702
Anxiety Disorder	0.0%	0.0%	0.0%	-
Cerebrovascular Disease	4.0%	3.8%	4.1%	0.831
Acute Confusional State	2.1%	1.7%	2.3%	0.789

Psychosis	1.3%	1.7%	1.1%	0.495
Myelopathy	0.5%	0.0%	0.7%	0.324
Movement Disorder	0.4%	0.8%	0.2%	0.213
Aseptic Meningitis	0.5%	0.8%	0.4%	0.587
Demyelinating Syndrome	0.5%	0.0%	0.7%	0.324
Polyneuropathy	1.3%	1.7%	1.1%	0.495
Mononeuropathy^β	1.6%	0.0%	2.3%	0.013
Cranial Neuropathy	1.3%	3.0%	0.5%	0.010
Acute Inflammatory Demyelinating Polyneuropathy	0.1%	0.0%	0.2%	1.000
Autonomic Disorder	0.3%	0.0%	0.4%	1.000
Plexopathy	0.0%	0.0%	0.0%	-
Myasthenia Gravis	0.3%	0.0%	0.4%	1.000
Any Central Neurologic Manifestation ^α	16.0%	14.4%	16.7%	0.414
Any Peripheral Neurologic Manifestation ^δ	4.3%	4.6%	4.1%	0.741
Any Neurological Manifestation	19.3%	17.7%	19.9%	0.478

Significant covariates are in **bold** (p<0.05).

p-values: Chi-squared test or Fisher's exact test.

^αACR NPSLE manifestations with onset within 10 years of SLE diagnosis and still present within the enrolment window or occurred subsequently; no 'exclusions'; not one of the NPSLE manifestations identified by Ainiola et al. (1).

^βMononeuropathy excluded from further analysis as no events observed in anti-KIF20B+ group and was non-significant in subsequent logistic regression analysis.

^αCentral Neurologic Manifestations: aseptic meningitis, cerebrovascular disease, cognitive dysfunction, headache, movement disorder, seizures, acute confusional state, anxiety disorder, mood disorder, psychosis, demyelinating syndrome, myelopathy.

^δPeripheral Neurologic Manifestations: autonomic disorder, mononeuropathy, cranial neuropathy, plexopathy, polyneuropathy, acute inflammatory demyelinating polyradiculoneuropathy, myasthenia gravis.

1. Ainiola H, Hietaharju A, Loukkola J, Peltola J, Korpela M, Metsänoja R, et al. Validity of the New American College of Rheumatology Criteria for Neuropsychiatric Lupus Syndromes: A Population-Based Evaluation. 2001 [cited 2023 Jan 2]; Available from: <https://onlinelibrary.wiley.com/doi/10.1002/1529-0131>

Supplemental Table 3: Clinical and demographic characteristics for SLE patients with CN						
	Anti-KIF20B Testing	Age at Disease Onset	CN Affected	SLEDAI-2K at Enrollment	Other SLE Manifestations	Other NPSLE Manifestations
Patient 1	Negative	42	Trochlear, abducens	0	Serositis, arthritis, hematologic, immunologic, ANA, dsDNA, Ro60, Ro52, La	Aseptic meningitis, stroke, polyneuropathy, autonomic disorder, tension headache, migraine headache

Patient 2	Positive	34	Vestibulo-cochlear	0	Oral ulcers, photosensitive rash, arthritis, hematologic, immunologic, ANA positive, dsDNA, PM/Scl	Mood disorder with depressive features
Patient 3	Positive	19	Facial	2	Arthritis, hematologic, immunologic, ANA, hypocomplementemic	Tension headache, migraine headache
Patient 4	Positive	45	Abducens, glossopharyngeal	8	Malar rash, arthritis, hematologic, immunologic, ANA, DSDNA, Ribosomal P	Aseptic meningitis, acute confusional state, cognitive dysfunction
Patient 5	Positive	20	Optic	18	Arthritis, renal, hematologic, ANA, vasculitis, hypocomplementemia, B2GP1, DSDNA, Histone, Ribosomal P, Smith, RNP, PCNA	Migraine headache, tension headache, mood disorder with depressive features
Patient 6	Negative	54	Abducens, vestibulo-cochlear	4	Arthritis, photosensitivity, Immunologic, ANA, hypocomplementemia, DSDNA, Ribosomal P, PCNA	
Patient 7	Positive	45	Optic	0	Malar, serositis, arthritis, hematologic, immunologic, ANA, DSDNA, histone	
Patient 8	Negative	37	Vestibulo-cochlear	2	Malar, photosensitive, hematologic, ANA, DNA binding, Ro 60	Autonomic disorder, migraine headache
Patient 9	Positive	26	Trigeminal	0	Arthritis, hematologic, immunologic, ANA, DSDNA, Histone, Jo-1, Ribosomal P, Smith, RNP, Scl-70, PM/Scl, CENP-B, PCNA, Ro52, Ro60, La	
Patient 10	Positive	38	Facial	2	Malar rash, oral ulcers, renal, hematologic, immunologic, ANA, DNA binding, DSDNA, Ribosomal P	Major depressive like episode, migraine headache

Supplemental Figure 1: Kaplan-Meier survival curve for anti-KIF20B positivity and cranial neuropathy within 5 years of follow-up

Supplemental Figure 2: Kaplan-Meier survival curve for cranial neuropathy and anti-KIF20B positivity and cranial neuropathy within all follow-up visits