

Supplementary Materials

Supplemental Table S1: Cohort of anti-OJ positive samples (n=39) detected by line immunoassay (LIA)*

Sample No.	Sample ID	cN-1A	Ro52	OJ	EJ	PL-12	PL-7	SRP	Jo-1	PM75	PM100	Ku	SAE1	NXP2	MDA5	TIF1 γ	Mi-2 β	Mi-2 α
1	X294			14				11										
2	X343		48	65							13							
3	X271			15														
4	X379			11														
5	X709			42												90		
6	X410			13														
7	X299			18		11												
8	X446			25														
9	X794			23														
10	X566		21	12						15						14		
11	X856			11														
12	X331			48														
13	X669			14								17						
14	X286			32														
15	X648			37														
16	X627			20														
17	X391	48		13			34					25						
18	X370			11														
19	X325			12						15								
20	X870			15														
21	X967			16														
22	X873			13														
23	X044			51														
24	X393			29								10						
25	X657			86														
26	X141			18														
27	A382			12														
28	X003			17														
29	X671			26				23										
30	X757		82	18								10		110				
31	X291			11								11						
32	X063			16														
33	X029	45		12														
34	X556			30														
35	X077			18														
36	X007			11														
37	X061			11														
38	X001			43														
39	X002			15														

* **LIA:** Results obtained from Euroimmun Myositis Profile 3 with EUROBlot Master 44, EUROLIne Scanner and software expressed as density units. Reference range 0-10 density units.

Supplemental Figure S1. Amino acid sequence alignment of human KARS and IARS proteins

sp Q15046 SYK_HUMAN	-----	0
sp P41252 SYIC_HUMAN	MLQQVPENINFPAAEEKILEFWTEFNCFQCECLKQSKHKPKFTFYDGPFFATGLPHYGHIL	60
sp Q15046 SYK_HUMAN	-----	0
sp P41252 SYIC_HUMAN	AGTIKDIVTRYAHQSGFHVDRRFQWDCGLPVEYEIDKTLGIRGPEVAKMGITEYNNQC	120
sp Q15046 SYK_HUMAN	-----	0
sp P41252 SYIC_HUMAN	RAIVMRYSAEWKSTVSRLGRWIDFDNDYKTLYPQFMESVWWVFKQLYDKGLVYRGVKVMP	180
sp Q15046 SYK_HUMAN	-----	0
sp P41252 SYIC_HUMAN	FSTACNTPLSNFESHQNYKDVQDPSVFVTFPLEEDETIVSLVAWTTTPWTLPNLAVCVNP	240
sp Q15046 SYK_HUMAN	-----	0
sp P41252 SYIC_HUMAN	EMQYVKIKDVARGRLILMEARLSALYKLESDEILERFPGAYLKGKKYRPLFDYFLCK	300
sp Q15046 SYK_HUMAN	-----	0
sp P41252 SYIC_HUMAN	ENGAFIVLVNDYVKEEGTGVVHQAPYFGAEDYRCMDFNIIKRDLSLPCVVDASGCFIT	360
sp Q15046 SYK_HUMAN	-----MAAYQAAEVKVD	12
sp P41252 SYIC_HUMAN	EVTDFAGQYVKDADKSIIRTLKEQGRLVATFTFHSYPFCWRSPTLIYKAVPSWFVRVE	420
sp Q15046 SYK_HUMAN	GSEPRLS-KNEL-----KRRLLAEK-----K-V	33
sp P41252 SYIC_HUMAN	NMVDQLLRNNDLCYVWPVLVREKRFGNMQRDARDWTISRNYWGTPILWVSDDFEFVVC	480
sp Q15046 SYK_HUMAN	AEKEAKQKELSEKQLSQATAAATNHT---TDNSVGPPEESVD-PNQQYKIRSQAIHCL--	87
sp P41252 SYIC_HUMAN	IGSVAELEELSGAKTSDLRHRESVDHLTIPSRCEKSSLHRISEVFTDWFESGSMFYAVHY	540
sp Q15046 SYK_HUMAN	----LVNCGEPYTHKHVDISLTDIFIQYSHLQPEDHLIDITLKVAGRIHAKRASGGKLI	143
sp P41252 SYIC_HUMAN	PFENRREFEAPFADDAIA-----EGIDQTRGWEYLLVL--ATA-----LFGQPP	583
sp Q15046 SYK_HUMAN	SYDLRGEVVKIQ----VMANSRNYKSEEEETHINNLRRGDIIGVQGNPGKTKKCELSI	198
sp P41252 SYIC_HUMAN	EKNVIVNELVVASDGQKMSKRKKNPDPVSIHQ---K-----YSADAL	623
sp Q15046 SYK_HUMAN	IPYETLLSLCLHMLPHHGLKDKETRYRQRYLLIINDFVROKRIIRSKI-----	250
sp P41252 SYIC_HUMAN	RLVIL--NSGVV-RAENLR-----KEGVRVQLKQVLPWYNAYRDLQNVLRLOKEEEI	676
sp Q15046 SYK_HUMAN	-----ITYIRSFIL-DELSFRIETPMNNII---E-----	275
sp P41252 SYIC_HUMAN	EFLYNENTVRESNITDRWILSDMQSLIEFEETEMAAAYRLYTVMRLVKFVDILTNNWVR	736
sp Q15046 SYK_HUMAN	-----CGAVAKFEITYHNEID-----	291
sp P41252 SYIC_HUMAN	MNRRRLKGENGMEDCVMALETLSVLLSLCLMAFYTPFELTELMYQNLKVLIDPVSVDK	796
sp Q15046 SYK_HUMAN	---MN---LYMRAPSLYHK--MLVGGIDRVYELIQRQRNEG-----	326
sp P41252 SYIC_HUMAN	DTLSIHVLMPLRVREELIDKTESAVSQMSVIELGRVIRDRKTIPIKYPLKEIVVIHQD	856
sp Q15046 SYK_HUMAN	-----IDITHNFEFTTCFYMAYADYHDLFEI	353
sp P41252 SYIC_HUMAN	PEALKDIKSLEKYIEELNVRKVTLSIDKNKYGRIRAEEDHMLVGLKRL-KGAFKAVTTS	915
sp Q15046 SYK_HUMAN	TEKM-----VSCMVKIIITGSYKYTYHDPGEQAYDVDFTPPFRIRINMVE	398
sp P41252 SYIC_HUMAN	IKQLSSEELQFQKTGTIVVEGHELD-DEDIRLMYTFQATGTGAQFEAHSDAQALVL--	972
sp Q15046 SYK_HUMAN	ELEKALGMKLEETNLEETEETRKILDDICVAKAVEG---PPRTTARLLDKLVGEFEVT	455
sp P41252 SYIC_HUMAN	-----LDVTEQDSMVDEGMAREVINRQKLR-KKQNLVETDEIVVYKAKSESTYLSV	1025
sp Q15046 SYK_HUMAN	CI-NPTFICDHPQIMSPAKWHRSKEGLTERFELEFVMKKEICNAYTESNDPMRQRIIFEE	514
sp P41252 SYIC_HUMAN	IESHTETIF--TTIKAPIKPYVPSPSDK-----VLIOEKQOLK----GSELEIT	1068
sp Q15046 SYK_HUMAN	QAKAKAGDDEAMFIDENFCIALE-----YGLPPTAGWGMGIDR	553
sp P41252 SYIC_HUMAN	LTRGSSSLPGPACAYVNLNLCANGSEQGVLLLENPKGDNRLDLLKLSVVTSIFVKNTE	1128
sp Q15046 SYK_HUMAN	VAMGLTDSNNI--KEVILFP-----AMKPEDK--	578
sp P41252 SYIC_HUMAN	LAVFIDEETIQNQTDLSLSGKTLCVTAGSAPSLINSSSTLLCQYINLQLLNAKQOECLM	1188
sp Q15046 SYK_HUMAN	-----KENVATDTTLE	589
sp P41252 SYIC_HUMAN	GTVGTLLLENPLGQNLTHQGLLYEAAKVFGRLSRKRLKFLNETQTQEITEDI-PVKTLN	1247
sp Q15046 SYK_HUMAN	STTVGTSV-----	597
sp P41252 SYIC_HUMAN	MRTVYVSULPTTADF	1262

Legend: Multiple alignment was carried out by Clustal Omega (RRID:SCR_001591). Red background with white letters shows strictly conserved residues; yellow background indicates conservative substitutions; grey background shows semi-conservative substitutions. Gaps are indicated as dashes. Percent identity and similarity of human KARS to IARS are 8.61% and 15.73% respectively.

Supplement Figure S2: Correlations of HEp-2 IIF Staining Patterns with Levels of Anti-IARS and Anti-KARS Autoantibodies Measured by PMAT

