



Autoantibodies in Diseases of Neuromuscular Junction




Myasthenia gravis

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Lambert-Eaton myastenic syndrome

Neuromyotonia

Morvan's syndrome

- Diagnostic marker with pathogenetic significance
 - Diagnostic marker for indicated disease
 - Indicative autoantibody
 - Occasionally associated autoantibodies with marker function for other diseases
 - Autoantibodies, which may be found randomly associated but without disease specificity and without any diagnostic significance for the disease mentioned.
- ▶ Cited **literature** is marked with red numbers and linked with the authors given at the end of the document. By clicking the hand symbol () one returns to the top of the table.
- ▶ **Autoantibodies** cited in the following tables are linked with their respective descriptions.



Autoantibodies in Diseases of Neuromuscular Junction



Myasthenia gravis

Autoantibodies	Sens [%]	Spec [%]	Disease associations
● Acetylcholine receptor (AChRm)	50 - 90	high	
● MuSK *	< 10	high	15
● Lrp4	< 10	high	
● Agrin	< 10	high	26, 27
● Potassium channel Kv1.4	12 - 15	high	
● Collagen Q	3	high	28
● Ryanodin receptor (RyR)	15 - 20	high	thymoma
● Titin	30	high	thymoma
● Dihydropyridin receptor (Ca _v 1.1)	37	high	thymoma
● TRPC3 channels	36	high	thymoma
● Gravin (AKAP-12)	31	high	4
● Rapsyn	low	medium	1
● Skeletal muscle (IIFT)	80	medium	
● Aquaporin-4	low	low	NMO 3, 7, 8, 13, 19
● CASPR2	low	low	NM, MS 19, 20
● Actin	< 5	low	AH 14, 17, 21, 22
● α-Actinin	< 25	low	AH 14, 17, 21, 22
● Filamin	< 10	low	9, 23
● Heat shock protein 70 (Hsp70)	< 20	low	6, 12
● Heat shock Protein HSC71	< 20	low	12
● Interferon-α2	< 20	low	2, 5, 10, 11, 16
● Interferon-α8	< 20	low	2, 5, 10, 11, 16
● Interferon-ω	< 20	low	2, 5, 10, 11
● Interleukin-12	< 20	low	5, 10, 11, 16, 18, 24, 25
● Myosin	< 50	low	14, 17, 21, 22
● Tropomyosin	< 30	low	17, 23
● Troponin	< 30	low	17
● Vinculin	< 10	low	23

AH: autoimmune hepatitis MS: Morvan's-syndrome NM: neuromyotonia
NMO: neuromyelitis optica

* an antigen equivalent to MuSK has been described and named protein p110 ¹⁵

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Lambert-Eaton-Syndrome

Autoantibodies	Sens [%]	Spec [%]	Disease associations
● Calcium channel (Overview)	90 - 100	high	SCLC (60%), LE, CD, SN, DA
● Calcium channel Cav2.1 (type P/Q)	90 - 100 ^{*1}	high	SN, DA 8, 10
● Calcium channel Cav2.2 (type N)	33 - 49 ^{*1}	high	DA 7, 8, 10
● Calcium channel Cav1.1 (type L)	case reports	unidentified	1, 5
● Calcium channel subunit β	23 - 55	unidentified	14, 15, 24
● SOX1 ^{*2}	bis 67	high (SCLC)	marker for SCLC ²¹ 16, 20
● Synaptotagmin 1	30 ^{*3}	unidentified	2, 17
● Acetylcholine receptor (AChRm)	case reports		MG/LEMS overlap 6, 11, 12
● M1 mACholine receptor	70 ^{*4}	unidentified	18, 19
● Glutamate decarboxylase (GAD)	35	unidentified	4
● IA-2 (tyrosine phosphatase)	21	unidentified	4
● Hu	case reports ^{*5}		3, 9, 13, 22
● CV2/CRMP5	5	unidentified	16
● PCA-2	case reports		23

CD: cerebellar degeneration DA: dysautonomia LE: limbic encephalitis
 LEMS: Lambert-Eaton myastenic syndrome MG: myasthenia gravis
 SCLC: small cell lung carcinoma SN: sensory neuropathy

- ^{*1} less frequent in paraneoplastic forms with SCLC
- ^{*2} antibodies may also crossreact with SOX2, SOX3, SOX21
- ^{*3} not found in all patient populations discribed ²
- ^{*4} existence also in anti-VGCC negative LEMS-patients 18, 19
- ^{*5} 9 % see Mason et al. (1997) ⁹

see also: Autoantibodies in Paraneoplastic Neuropathies

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Autoantibodies in Diseases of Neuromuscular Junction



Neuromyotonia

Autoantibodies	Sens [%]	Spec [%]	Disease associations
● Potassium channel complex* ¹	54 ¹³ - 95	high * ²	thymoma , SCLC, DA 3, 4, 5
● Potassium channel Kv1.1 * ³	< 3		4, 7, 8
● Potassium channel Kv1.2 * ³	< 3		4, 7, 8
● Potassium channel Kv1.6 * ³	< 3		4, 7, 8
● CASPR2	high	high * ²	7, 9, 14
● LGI1	low	high * ²	2, 7
● Tag-1/Contactin 2	low	high * ²	7
● Acetylcholine receptor (muscular)	14 ¹³		thymoma 6, 13
● Acetylcholine receptor (ganglionic)	14 ¹³		associated tumors * ⁴ 12, 13
● MuSK	case reports		11a
● Ryanodin receptor (RyR)	case reports		thymoma 10
● Amphiphysin	case reports		paraneoplasia 11
● Glutamate decarboxylase (GAD)	case reports		thymoma 1

DA: dysautonomia SCLC: small cell lung carcinoma

- *¹ In the majority of patients the antibodies do not react with the pore forming proteins Kv1.1, 1.2, 1.6 but with the associated channel proteins CASPR2, LGI1, Tag-1 (channel complex).
- *² Valid for neuromyotonia, Morvan's syndrome, and limbic encephalitis.
- *³ Antibodies demonstrated by IIFT in transfected, the corresponding channel protein expressing culture cells (4, 8); the results could not be confirmed regularly (7).
- *⁴ Thymoma (anti-CRMP5 positive), SCLC (anti-CRMP5 positive), carcinoma of the lung (anti-Amphiphysin positive), SCLC (ANNA positive) (13).

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Autoantibodies in Diseases of Neuromuscular Junction



Morvan's syndrome

Autoantibodies	Sens [%]	Spec [%]	Disease associations
● Potassium channel complex ^{*1}	40 - 80	high ^{*3}	LE, NM, EP, thymoma 1, 6, 7
● Potassium channel Kv1.1 ^{*2}	< 3		4
● Potassium channel Kv1.2 ^{*2}	< 3		4
● Potassium channel Kv1.6 ^{*2}	< 3		4
● CASPR2	high	high ^{*3}	LE, NM, EP 9
● LGI1	low	high ^{*3}	LE, NM, EP 5
● Tag-1/Contactin 2	low	high ^{*3}	8
● Acetylcholine receptor (AChRm)	case reports		thymoma, MG/MS 2, 3, 6
● Titin	case reports		thymoma 6
● Skeletal muscle (IIFT)	case reports		thymoma 6
● Calcium channel Cav2.2 (type N)	case reports		6

LE: limbic encephalitis NM: neuromyotonia EP: epilepsy, seizures

MG/MS: overlap syndrome of myasthenia gravis and anti-AChR, anti-MuSK, anti-VGKC resp.

^{*1} In the majority of patients the antibodies do not react with the pore forming proteins Kv1.1, 1.2, 1.6 but with the associated channel proteins CASPR2, LGI1, Tag-1 (channel complex).

^{*2} Antibodies demonstrated by IIFT in transfected, the corresponding channel protein expressing culture cells (4); the results could not be confirmed regularly (see literature: table neuromyotonia, 7).

^{*3} Valid for neuromyotonia, Morvan's syndrome, and limbic encephalitis.

see also: Autoantibodies in paraneoplastic neuropathies

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


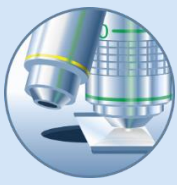
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Abbreviations

Sens	sensitivity
Spec	specificity
CASPR2	contactin-assoziated protein-2
CV2/CRMP5	collapsing response mediator protein 5
Hu	abbreviation of the name from a patient
IA2	islet cell antigen 2
IIFT	indirect immunofluoreszenze test
GAD	glutamate decarboxylase
LGI1	leucine-rich glioma inactivated 1
Lrp4	low density lipoprotein receptor-related protein 4
M1 mAChR	M1 muscarinic acetylcholine receptor
MuSK	muscle specific tyrosine kinase
PCA-2	pukinje cell antibody Typ 2
SOX1	sex determining region y-box 1
Tag-1/Contactin 2	transient axonal glycoprotein 1
TRPC3	transient receptor potential canonical type 3
VGCC	voltage gated calcium channel
VGKC	voltage gated potassium channel

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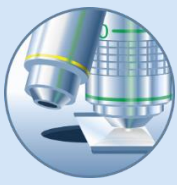
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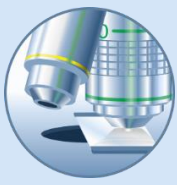


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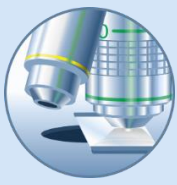
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