



Autoantikörper bei Erkrankungen der neuromuskulären Transmission



Myasthenia gravis

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Lambert-Eaton myasthenisches Syndrom

Neuromyotonie (Isaac-Syndrom)

Morvan-Syndrom

Literatur

- Diagnostischer Krankheitsmarker von pathogener Bedeutung
 - Diagnostischer Krankheitsmarker der Grunderkrankung
 - hinweisender Autoantikörper
 - Autoantikörper auch mit anderen Krankheiten assoziiert
 - Bei dem betreffenden Krankheitsbild ebenfalls anzutreffende, aber nicht krankheitsspezifische Autoantikörper. Keine diagnostische Bedeutung für die beschriebene Erkrankung.
- **Literaturzitate** sind mit roten Zahlen markiert und auf die Autoren am Ende des Dokuments verlinkt. Durch Anklicken des Handsymbols (☞) gelangt man an den Tabellenanfang zurück.
- Die in der Tabelle aufgeführten Autoantikörper sind mit ihrer Beschreibung verlinkt.



Autoantikörper bei Erkrankungen der neuromuskulären Transmission

Myasthenia gravis

Autoantikörper gegen	Sens [%]	Spez [%]	Krankheitsassoziation
● Acetylcholinrezeptor (AChRm)	50 - 90	hoch	
● MuSK *	< 10	hoch	15
● Lrp4	< 10	hoch	
● Agrin	< 10	hoch	26, 27
● Kaliumkanal Kv1.4	12 - 15	hoch	
● Kollagen Q	3	hoch	28
● Ryanodinrezeptor (RyR)	15 - 20	hoch	Thymom
● Titin	30	hoch	Thymom
● Dihydropyridinrezeptor (Ca _v 1.1)	37	hoch	Thymom
● TRPC3 Kanäle	36	hoch	Thymom
● Gravin (AKAP-12)	31	hoch	4
● Rapsyn	nieder	mittel	1
● Skelettmuskel (IIFT)	80	mittel	
● Aquaporin-4	nieder	nieder	NMO 3, 7, 8, 13, 19
● CASPR2	nieder	nieder	NM, MS 19, 20
● Actin	< 5	nieder	AH 14, 17, 21, 22
● α-Actinin	< 25	nieder	AH 14, 17, 21, 22
● Filamin	< 10	nieder	9, 23
● Heat shock Protein 70	< 20	nieder	6, 12
● Heat shock Protein HSC71	< 20	nieder	12
● Interferon-α2	< 20	nieder	2, 5, 10, 11, 16
● Interferon-α8	< 20	nieder	2, 5, 10, 11, 16
● Interferon-ω	< 20	nieder	2, 5, 10, 11
● Interleukin-12	< 20	nieder	5, 10, 11, 16, 18, 24, 25
● Myosin	< 50	nieder	14, 17, 21, 22
● Tropomyosin	< 30	nieder	17, 23
● Troponin	< 30	nieder	17
● Vinculin	< 10	nieder	23

AH: autoimmune Hepatitis MS: Morvan-Syndrome NM: Neuromyotonie
NMO: Neuromyelitis optica

* ein mit MuSK identisches Antigen wurde unter der Bezeichnung Protein p110 beschrieben 15

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Lambert-Eaton myasthenisches Syndrom

Autoantikörper gegen	Sens [%]	Spez [%]	Krankheitsassoziation
● Calciumkanäle (VGCC)	90 - 100	hoch	SCLC (60%), LE, CD, SN, DA
● Calciumkanal Ca _v 2.1 (P/Q-Typ)	90 - 100 ^{*1}	hoch	SN, DA 8, 10
● Calciumkanal Ca _v 2.2 (N-Typ)	33 - 49 ^{*1}	hoch	DA 7, 8, 10
● Calciumkanal Ca _v 1.1 (L-Typ)	Kasuistiken	unbekannt	1, 5
● Calciumkanal β-Untereinheit	23 - 55	unbekannt	14, 15, 24
● SOX1 ^{*2}	bis 67	hoch (SCLC)	Frühmarker für SCLC ²¹ 16, 20
● Synaptotagmin 1	30 ^{*3}	unbekannt	2, 17
● Acetylcholinrezeptor (AChRm)	Kasuistiken		MG/LEMS overlap 6, 11, 12
● M1 mACholinrezeptor	70 ^{*4}	unbekannt	18, 19
● Glutamatdecarboxylase (GAD)	35	unbekannt	4
● IA-2 (Thyrosinphosphatase)	21	unbekannt	4
● HuD	Kasuistiken ^{*5}		3, 9, 13, 22
● CV2/CRMP5	5	unbekannt	16
● PCA-2	Kasuistiken		23

CD: cerebellare Degeneration DA: Dysautonomie LE: limbische Enzephalitis
LEMS: Lambert-Eaton myasthenisches Syndrom MG: Myasthenia gravis
SCLC: kleinzelliges Lungenkarzinom SN: sensorische Neuropathie

^{*1} weniger häufig bei paraneoplastischen Formen mit SCLC

^{*2} Kreuzreaktion mit SOX2, SOX3, SOX21

^{*3} wurde nicht bei allen beschriebenen Patientenkollektiven gefunden ²

^{*4} Vorkommen auch bei anti-VGCC negativen LEMS-Patienten beschrieben 18, 19

^{*5} 9 % siehe Mason et al. (1997), ⁹

siehe: [Paraneoplastische Neuropathien](#)

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Neuromyotonie

Autoantikörper gegen	Sens [%]	Spez [%]	Krankheitsassoziation
● Kaliumkanalkomplex * ¹	54 ¹³ - 95	hoch * ²	Thymom , SCLC, DA 3, 4, 5
● Kaliumkanal Kv1.1 * ³	< 3		4, 7, 8
● Kaliumkanal Kv1.2 * ³	< 3		4, 7, 8
● Kaliumkanal Kv1.6 * ³	< 3		4, 7, 8
● CASPR2	hoch	hoch * ²	7, 9, 14
● LGI1	nieder	hoch * ²	2, 7
● Tag-1/Contactin 2	nieder	hoch * ²	7
● Acetylcholinrezeptor (muskulärer)	14 ¹³		Thymom 6, 13
● Acetylcholinrezeptor (ganglionärer)	14 ¹³		assoziierte Tumore * ⁴ 12, 13
● MuSK	Kasuistiken		11a
● Ryanodinrezeptor (RyR)	Kasuistiken		Thymom 10
● Amphiphysin	Kasuistiken		Paraneoplasie 11
● Glutamatdecarboxylase (GAD)	Kasuistiken		Thymom 1

DA: Dysautonomie SCLC: kleinzelliges Lungenkarzinom

- *¹ Die Antikörper richten sich in den meisten Fällen nicht gegen die Kanalporen-bildenden (Kv1.1, 1.2, 1.6) sondern gegen die assoziierten Proteine CASPR2, LGI-1, Tag-1 (Kanal-Komplex).
- *² Gültig für Neuromyotonie, Morvan-Syndrom und limbische Enzephalitis.
- *³ Autoantikörper nachgewiesen mittels IIFT an transfizierten, das entsprechende Kanalprotein exprimierenden Kulturzellen (4, 8); die Ergebnisse ließen sich nicht regelmäßig bestätigen (7).
- *⁴ Thymom (anti-CRMP5 positiv), SCLC (anti-CRMP5 positiv), Lungenkarzinom (anti-Amphiphysin positiv), SCLC (ANNA positiv) (13).

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Morvan-Syndrom

Autoantikörper gegen	Sens [%]	Spez [%]	Krankheitsassoziation
● Kaliumkanalkomplex * ¹	40 - 80	hoch * ³	LE, NM, EP, Thymom 1, 6, 7
● Kaliumkanal Kv1.1 * ²	< 3		4
● Kaliumkanal Kv1.2 * ²	< 3		4
● Kaliumkanal Kv1.6 * ²	< 3		4
● CASPR2	hoch	hoch * ³	LE, NM, EP 9
● LGI1	nieder	hoch * ³	LE, NM, EP 5
● Tag-1/Contactin 2	nieder	hoch * ³	8
● Acetylcholinrezeptor (AChRm)	Kasuistik		Thymom, MG/MS 2, 3, 6
● Titin	Kasuistik		Thymom 6
● Skelettmuskel (IIFT)	Kasuistik		Thymom 6
● Calciumkanal Ca _v 2.2 (N-Typ)	Kasuistik		6

LE: limbische Enzephalitis NM Neuromyotonie EP Epilepsie, Krampfanfälle

MG/MS: Überlappungssyndrom mit Myasthenia gravis und anti-AChR, anti-MuSK und anti-VGKC

*¹ Die Antikörper richten sich in den meisten Fällen nicht gegen die Kanalporen-bildenden (Kv1.1, 1.2, 1.6) sondern gegen die assoziierten Proteine CASPR2, LGI-1, Tag-1 (Kanal-Komplex).

*² Autoantikörper nachgewiesen mittels IIFT an transfizierten, das entsprechende Kanalprotein exprimierenden Kulturzellen (4); die Ergebnisse ließen sich nicht regelmäßig bestätigen (vergl. Literatur: Neuromyotonie 7).

*³ Gültig für Neuromyotonie, Morvan-Syndrom und limbische Enzephalitis.

siehe: [Autoantikörper bei paraneoplastische Neuropathien](#)

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


Autoantikörper bei Erkrankungen der neuromuskulären Transmission



Abkürzungen

Sens	Sensitivität
Spez	Spezifität
CASPR2	contactin-assoziiated protein-2
CV2/CRMP5	collapsing response mediator protein 5
Hu	Abkürzung eines Patientennamens
IA2	islet cell antigen 2
IIFT	indirekter Immunfluoreszenztest
GAD	Glutamatdecarboxylase
LGI1	leucine-rich glioma inactivated 1
Lrp4	low density lipoprotein receptor-related protein 4
M1 mAChR	M1 muskarinischer Acetylcholinrezeptor
MuSK	mukelspezifische Tyrosinkinase
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 (Calciumkanal)
VGKC	voltage gated potassium channel (Kaliumkanal)

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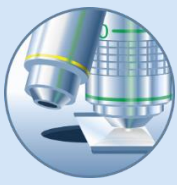


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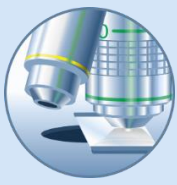
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