Mathias Gautel

List of Publications by Year
in descending order

120	17,099	18436	19690
papers	citations	62 h-index	117 g-index

127	127	127	20427
all docs	docs citations	times ranked	citing authors

#	Article	IF	Citations
1	The spectrum of neurodevelopmental, neuromuscular and neurodegenerative disorders due to defective autophagy. Autophagy, 2022, 18, 496-517.	4.3	18
2	Structures from intact myofibrils reveal mechanism of thin filament regulation through nebulin. Science, 2022, 375, eabn1934.	6.0	69
3	High Throughput Screen Identifies Small Molecule Effectors That Modulate Thin Filament Activation in Cardiac Muscle. ACS Chemical Biology, 2021, 16, 225-235.	1.6	7
4	When is an obscurin variant pathogenic? The impact of Arg4344Gln and Arg4444Trp variants on proteinâ€“protein interactions and protein stability. Human Molecular Genetics, 2021, 30, 1131-1141.	1.4	16
5	The molecular basis for sarcomere organization in vertebrate skeletal muscle. Cell, 2021, 184, 2135-2150.e13.	13.5	99
6	Order from disorder in the sarcomere: FATZ forms a fuzzy but tight complex and phase-separated condensates with Î±-actinin. Science Advances, 2021, 7, .	4.7	15
7	Molecular noise filtering in the Î²-adrenergic signaling network by phospholamban pentamers. Cell Reports, 2021, 36, 109448.	2.9	5
8	The molecular basis for sarcomere organization in vertebrate skeletal muscle. Microscopy and Microanalysis, 2021, 27, 2832-2835.	0.2	1
9	Phosphorylation at Serines 157 and 161 Is Necessary for Preserving Cardiac Expression Level and Functions of Sarcomeric Z-Disc Protein Telethonin. Frontiers in Physiology, 2021, 12, 732020.	1.3	1
10	Sub-diffraction error mapping for localisation microscopy images. Nature Communications, 2021, 12, 5611.	5.8	14
11	Myopalladin knockout mice develop cardiac dilation and show a maladaptive response to mechanical pressure overload. ELife, 2021, 10, .	2.8	12
12	Making sense of missense variants in TTN-related congenital myopathies. Acta Neuropathologica, 2021, 141, 431-453.	3.9	34
13	The Axial Alignment of Titin on the Muscle Thick Filament Supports Its Role as a Molecular Ruler. Journal of Molecular Biology, 2020, 432, 4815-4829.	2.0	21
14	Cardiac myosin regulatory light chain kinase modulates cardiac contractility by phosphorylating both myosin regulatory light chain and troponin I. Journal of Biological Chemistry, 2020, 295, 4398-4410.	1.6	16
15	Congenital myopathies: disorders of excitationâ€“contraction coupling and muscle contraction. Nature Reviews Neurology, 2018, 14, 151-167.	4.9	212
16	Artifact-free high-density localization microscopy analysis. Nature Methods, 2018, 15, 689-692.	9.0	79
17	219th ENMC International Workshop Titinopathies International database of titin mutations and phenotypes, Heemskerk, The Netherlands, 29 Aprilâ€“1 May 2016. Neuromuscular Disorders, 2017, 27, 396-407.	0.3	29
18	Binding of Myomesin to Obscurin-Like-1 at the Muscle M-Band Provides a Strategy for Isoform-Specific Mechanical Protection. Structure, 2017, 25, 107-120.	1.6	25

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19	Autopsy findings in EPG5â€related Vici syndrome with antenatal onset. American Journal of Medical Genetics, Part A, 2017, 173, 2522-2527.	0.7	6
20	TITINdbâ€”a computational tool to assess titinâ€™s role as a disease gene. Bioinformatics, 2017, 33, 3482-3485.	1.8	34
21	Current and future therapeutic approaches to the congenital myopathies. Seminars in Cell and Developmental Biology, 2017, 64, 191-200.	2.3	29
22	Increasing evidence of mechanical force as a functional regulator in smooth muscle myosin light chain kinase. ELife, 2017, 6, .	2.8	15
23	Combination of Whole Genome Sequencing, Linkage, and Functional Studies Implicates a Missense Mutation in Titin as a Cause of Autosomal Dominant Cardiomyopathy With Features of Left Ventricular Noncompaction. Circulation: Cardiovascular Genetics, 2016, 9, 426-435.	5.1	67
24	Reply: Aberrant splicing induced by the most commonEPG5mutation in an individual with Vici syndrome. Brain, 2016, 139, e53-e53.	3.7	4
25	Vici syndrome: a review. Orphanet Journal of Rare Diseases, 2016, 11, 21.	1.2	55
26	The sarcomeric cytoskeleton: from molecules to motion. Journal of Experimental Biology, 2016, 219, 135-145.	0.8	188
27	EPG5-related Vici syndrome: a paradigm of neurodevelopmental disorders with defective autophagy. Brain, 2016, 139, 765-781.	3.7	99
28	Epigenetic changes as a common trigger of muscle weakness in congenital myopathies. Human Molecular Genetics, 2015, 24, 4636-4647.	1.4	44
29	Titin ruler hypothesis not refuted. Proceedings of the National Academy of Sciences of the United States of America, 2015, 112, E1172-E1172.	3.3	16
30	Binding partners of the kinase domains inDrosophilaobscurin and their effect on the structure of the flight muscle. Journal of Cell Science, 2015, 128, 3386-97.	1.2	24
31	The Crystal Structure of the Human Titin:Obscurin Complex Reveals a Conserved yet Specific Muscle M-Band Zipper Module. Journal of Molecular Biology, 2015, 427, 718-736.	2.0	20
32	Solution NMR assignment of the heavy chain complex of the human cardiac myosin regulatory light chain. Biomolecular NMR Assignments, 2015, 9, 51-53.	0.4	1
33	Pathogenic Mechanisms in Centronuclear Myopathies. Frontiers in Aging Neuroscience, 2014, 6, 339.	1.7	89
34	Phosphoregulation of the Titin-cap Protein Telethonin in Cardiac Myocytes. Journal of Biological Chemistry, 2014, 289, 1282-1293.	1.6	32
35	The Structure and Regulation of Human Muscle Î±-Actinin. Cell, 2014, 159, 1447-1460.	13.5	178
36	Clinical utility gene card for: Vici Syndrome. European Journal of Human Genetics, 2014, 22, 435-435.	1.4	13

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37	Reply: Hereditary myopathy with early respiratory failure is caused by mutations in the titin FN3 119 domain. Brain, 2014, 137, e279-e279.	3.7	13
38	Myosin binding protein-C activates thin filaments and inhibits thick filaments in heart muscle cells. Proceedings of the National Academy of Sciences of the United States of America, 2014, 111, 18763-18768.	3.3	103
39	Recessive TTN truncating mutations define novel forms of core myopathy with heart disease. Human Molecular Genetics, 2014, 23, 980-991.	1.4	149
40	MuRF1 activity is present in cardiac mitochondria and regulates reactive oxygen species production in vivo. Journal of Bioenergetics and Biomembranes, 2014, 46, 173-187.	1.0	23
41	Diggināâ€² on U(biquitin): A Novel Method for the Identification of Physiological E3 Ubiquitin Ligase Substrates. Cell Biochemistry and Biophysics, 2013, 67, 127-138.	0.9	15
42	Recessive mutations in EPG5 cause Vici syndrome, a multisystem disorder with defective autophagy. Nature Genetics, 2013, 45, 83-87.	9.4	231
43	Guidelines for the use and interpretation of assays for monitoring autophagy. Autophagy, 2012, 8, 445-544.	4.3	3,122
44	The sarcomeric cytoskeleton as a target for pharmacological intervention. Current Opinion in Pharmacology, 2012, 12, 347-354.	1.7	27
45	Developmental regulation of MURF E3 ubiquitin ligases in skeletal muscle. Journal of Muscle Research and Cell Motility, 2012, 33, 107-122.	0.9	46
46	Structure, interactions and function of the N-terminus of cardiac myosin binding protein C (MyBP-C): who does what, with what, and to whom?. Journal of Muscle Research and Cell Motility, 2012, 33, 83-94.	0.9	85
47	Introducing a series of topical special issues of the Journal of Muscle Research and Cell Motility. Journal of Muscle Research and Cell Motility, 2012, 33, 1-3.	0.9	0
48	A Conditional Gating Mechanism Assures the Integrity of the Molecular Force-Sensor Titin Kinase. Biophysical Journal, 2011, 101, 1978-1986.	0.2	20
49	Developmental regulation of MURF ubiquitin ligases and autophagy proteins nbr1, p62/SQSTM1 and LC3 during cardiac myofibril assembly and turnover. Developmental Biology, 2011, 351, 46-61.	0.9	57
50	Transcriptional mechanisms regulating skeletal muscle differentiation, growth and homeostasis. Nature Reviews Molecular Cell Biology, 2011, 12, 349-361.	16.1	570
51	Preferential skeletal muscle myosin loss in response to mechanical silencing in a novel rat intensive care unit model: underlying mechanisms. Journal of Physiology, 2011, 589, 2007-2026.	1.3	112
52	The sarcomeric cytoskeleton: who picks up the strain?. Current Opinion in Cell Biology, 2011, 23, 39-46.	2.6	163
53	Cytoskeletal protein kinases: titin and its relations in mechanosensing. Pflugers Archiv European Journal of Physiology, 2011, 462, 119-134.	1.3	111
54	Structure and Interactions of Myosin-binding Protein C Domain C0. Journal of Biological Chemistry, 2011, 286, 12650-12658.	1.6	114

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55	Editorial June 2010. Journal of Muscle Research and Cell Motility, 2010, 31, 1-1.	0.9	0
56	Gett'N-WASP Stripes. Science, 2010, 330, 1491-1492.	6.0	4
57	Structural insight into M-band assembly and mechanics from the titin-obscurin-like-1 complex. Proceedings of the National Academy of Sciences of the United States of America, 2010, 107, 2908-2913.	3.3	60
58	Distinct Sarcomeric Substrates Are Responsible for Protein Kinase D-mediated Regulation of Cardiac Myofilament Ca2+ Sensitivity and Cross-bridge Cycling. Journal of Biological Chemistry, 2010, 285, 5674-5682.	1.6	96
59	Interactions with LC3 and polyubiquitin chains link nbr1 to autophagic protein turnover. FEBS Letters, 2009, 583, 1846-1852.	1.3	78
60	Comparing Proteins by Their Unfolding Pattern. Biophysical Journal, 2008, 95, 426-434.	0.2	71
61	Myosin Binding Protein C Positioned to Play a Key Role in Regulation of Muscle Contraction: Structure and Interactions of Domain C1. Journal of Molecular Biology, 2008, 384, 615-630.	2.0	86
62	Mechanoenzymatics of titin kinase. Proceedings of the National Academy of Sciences of the United States of America, 2008, 105, 13385-13390.	3.3	311
63	Interactions with titin and myomesin target obscurin and obscurin-like 1 to the M-band â€“ implications for hereditary myopathies. Journal of Cell Science, 2008, 121, 1841-1851.	1.2	168
64	The Sarcomere and Sarcomerogenesis. Advances in Experimental Medicine and Biology, 2008, 642, 1-14.	0.8	81
65	The Sarcomere and the Nucleus: Functional Links to Hypertrophy, Atrophy and Sarcopenia. Advances in Experimental Medicine and Biology, 2008, 642, 176-191.	0.8	40
66	Dissecting the N-terminal Myosin Binding Site of Human Cardiac Myosin-binding Protein C. Journal of Biological Chemistry, 2007, 282, 9204-9215.	1.6	69
67	Rigid Conformation of an Immunoglobulin Domain Tandem Repeat in the A-band of the Elastic Muscle Protein Titin. Journal of Molecular Biology, 2007, 371, 469-480.	2.0	26
68	C-terminal titin deletions cause a novel early-onset myopathy with fatal cardiomyopathy. Annals of Neurology, 2007, 61, 340-351.	2.8	209
69	Evidence for a dimeric assembly of two titin/telethonin complexes induced by the telethonin C-terminus. Journal of Structural Biology, 2006, 155, 239-250.	1.3	25
70	Palindromic assembly of the giant muscle protein titin in the sarcomeric Z-disk. Nature, 2006, 439, 229-233.	13.7	166
71	Complete human gene structure of obscurin: implications for isoform generation by differential splicing. Journal of Muscle Research and Cell Motility, 2006, 26, 427-434.	0.9	65
72	From A to Z and back? Multicompartment proteins in the sarcomere. Trends in Cell Biology, 2006, 16, 11-18.	3.6	163

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73	Targeted homozygous deletion of M-band titin in cardiomyocytes prevents sarcomere formation. Journal of Cell Science, 2006, 119, 4322-4331.	1.2	74
74	Activation of Myocardial Contraction by the N-Terminal Domains of Myosin Binding Protein-C. Circulation Research, 2006, 98, 1290-1298.	2.0	80
75	Crystal structures of human cardiac beta-myosin II S2-Â provide insight into the functional role of the S2 subfragment. Proceedings of the National Academy of Sciences of the United States of America, 2006, 103, 17713-17717.	3.3	97
76	Calpain 1-titin interactions concentrate calpain 1 in the Z-band edges and in the N2-line region within the skeletal myofibril. FEBS Journal, 2005, 272, 2578-2590.	2.2	69
77	The Kinase Domain of Titin Controls Muscle Gene Expression and Protein Turnover. Science, 2005, 308, 1599-1603.	6.0	524
78	The Crystal Structure of the Actin Binding Domain from Î±-Actinin in its Closed Conformation: Structural Insight into Phospholipid Regulation of Î±-Actinin. Journal of Molecular Biology, 2005, 348, 151-165.	2.0	66
79	Mechanically Induced Titin Kinase Activation Studied by Force-Probe Molecular Dynamics Simulations. Biophysical Journal, 2005, 88, 790-804.	0.2	195
80	Binding of Myosin Binding Protein-C to Myosin Subfragment S2 Affects Contractility Independent of a Tether Mechanism. Circulation Research, 2004, 95, 930-936.	2.0	71
81	Association of the Chaperone Î±B-crystallin with Titin in Heart Muscle. Journal of Biological Chemistry, 2004, 279, 7917-7924.	1.6	147
82	Protein Kinase D Is a Novel Mediator of Cardiac Troponin I Phosphorylation and Regulates Myofilament Function. Circulation Research, 2004, 95, 1091-1099.	2.0	135
83	Letter to the Editor: Sequence Specific Assignment of Domain C1 of the N-terminal Myosin-binding Site of Human Cardiac Myosin Binding Protein C (MyBP-C). Journal of Biomolecular NMR, 2004, 29, 431-432.	1.6	5
84	The Elasticity of Single Titin Molecules Using a Two-Bead Optical Tweezers Assay. Biophysical Journal, 2004, 87, 1112-1135.	0.2	89
85	Myofibrillar tightly bound calcium in skeletal muscle fibers: a possible role of this cation in titin strands aggregation. FEBS Letters, 2004, 556, 271-275.	1.3	8
86	Structure, Stability and Dynamics of the Central Domain of Cardiac Myosin Binding Protein C (MyBP-C): Implications for Multidomain Assembly and Causes for Cardiomyopathy. Journal of Molecular Biology, 2003, 329, 745-761.	2.0	40
87	Solution Scattering Suggests Cross-linking Function of Telethonin in the Complex with Titin. Journal of Biological Chemistry, 2003, 278, 2636-2644.	1.6	45
88	Transient association of titin and myosin with microtubules in nascent myofibrils directed by the MURF2 RING-finger protein. Journal of Cell Science, 2002, 115, 4469-4482.	1.2	131
89	PEVK Domain of Titin: An Entropic Spring with Actin-Binding Properties. Journal of Structural Biology, 2002, 137, 194-205.	1.3	179
90	The spectrin repeat: a structural platform for cytoskeletal protein assemblies. FEBS Letters, 2002, 513, 119-123.	1.3	249

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91	Sequence specific resonance assignment of the central domain of cardiac myosin binding protein C (MyBP-C). Journal of Biomolecular NMR, 2002, 22, 199-200.	1.6	4
92	Kinase recognition by calmodulin: modeling the interaction with the autoinhibitory region of human cardiac titin kinase11Edited by J. Thornton. Journal of Molecular Biology, 2001, 306, 81-95.	2.0	8
93	Early and selective disappearance of telethonin protein from the sarcomere in neurogenic atrophy. Journal of Muscle Research and Cell Motility, 2001, 22, 259-264.	0.9	18
94	Structural Evidence for a Possible Role of Reversible Disulphide Bridge Formation in the Elasticity of the Muscle Protein Titin. Structure, 2001, 9, 331-340.	1.6	80
95	Obscurin, a giant sarcomeric Rho guanine nucleotide exchange factor protein involved in sarcomere assembly. Journal of Cell Biology, 2001, 154, 123-136.	2.3	256
96	Myosin Binding Protein C, a Phosphorylation-Dependent Force Regulator in Muscle That Controls the Attachment of Myosin Heads by Its Interaction With Myosin S2. Circulation Research, 2000, 86, 51-58.	2.0	200
97	Characterization of muscle filamin isoforms suggests a possible role of ?-filamin/ABP-L in sarcomeric Z-disc formation. Cytoskeleton, 2000, 45, 149-162.	4.4	141
98	A Newly Created Splice Donor Site in Exon 25 of the MyBP-C Gene Is Responsible for Inherited Hypertrophic Cardiomyopathy With Incomplete Disease Penetrance. Circulation, 2000, 101, 1396-1402.	1.6	114
99	Unfolding Forces of Titin and Fibronectin Domains Directly Measured by AFM. Advances in Experimental Medicine and Biology, 2000, 481, 129-141.	0.8	71
100	I-Band Titin in Cardiac Muscle Is a Three-Element Molecular Spring and Is Critical for Maintaining Thin Filament Structure. Journal of Cell Biology, 1999, 146, 631-644.	2.3	228
101	cAPK-phosphorylation controls the interaction of the regulatory domain of cardiac myosin binding protein C with myosin-S2 in an on-off fashion. FEBS Letters, 1999, 453, 254-259.	1.3	169
102	Molecular Basis for Cross-Linking of Actin Filaments: Structure of the Î±-Actinin Rod. Cell, 1999, 98, 537-546.	13.5	237
103	Mutations in Î²-myosin S2 that cause familial hypertrophic cardiomyopathy (FHC) abolish the interaction with the regulatory domain of myosin-binding protein-C 1 1Edited by J. Karn. Journal of Molecular Biology, 1999, 286, 933-949.	2.0	221
104	A six-module human nebulin fragment bundles actin filaments and induces actin polymerization. Journal of Muscle Research and Cell Motility, 1998, 19, 225-235.	0.9	24
105	The Mechanical Stability of Immunoglobulin and Fibronectin III Domains in the Muscle Protein Titin Measured by Atomic Force Microscopy. Biophysical Journal, 1998, 75, 3008-3014.	0.2	302
106	Structural basis for activation of the titin kinase domain during myofibrillogenesis. Nature, 1998, 395, 863-869.	13.7	333
107	Two immunoglobulin-like domains of the Z-disc portion of titin interact in a conformation-dependent way with telethonin. FEBS Letters, 1998, 428, 111-114.	1.3	144
108	SH3 in muscles: solution structure of the SH3 domain from nebulin. Journal of Molecular Biology, 1998, 276, 189-202.	2.0	40

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109	The assembly of immunoglobulin-like modules in titin: implications for muscle elasticity. Journal of Molecular Biology, 1998, 284, 761-777.	2.0	78
110	Isoform Transitions of the Myosin Binding Protein C Family in Developing Human and Mouse Muscles. Circulation Research, 1998, 82, 124-129.	2.0	104
111	Reversible Unfolding of Individual Titin Immunoglobulin Domains by AFM. Science, 1997, 276, 1109-1112.	6.0	2,874
112	A survey of in situ sarcomere extension in mouse skeletal muscle. Journal of Muscle Research and Cell Motility, 1997, 18, 465-472.	0.9	22
113	Constitutive and Variable Regions of Z-disk Titin/Connectin in Myofibril Formation: A Dominant-negative Screen.. Cell Structure and Function, 1997, 22, 95-101.	0.5	36
114	A molecular map of titin/connectin elasticity reveals two different mechanisms acting in series. FEBS Letters, 1996, 385, 11-14.	1.3	100
115	Titin Domain Patterns Correlate with the Axial Disposition of Myosin at the End of the Thick Filament. Journal of Molecular Biology, 1996, 259, 896-903.	2.0	44
116	A Molecular Map of the Interactions between Titin and Myosin-Binding Protein C. Implications for Sarcomeric Assembly in Familial Hypertrophic Cardiomyopathy. FEBS Journal, 1996, 235, 317-323.	0.2	249
117	Assembly of the cardiac I-band region of titin/connectin: expression of the cardiac-specific regions and their structural relation to the elastic segments. Journal of Muscle Research and Cell Motility, 1996, 17, 449-461.	0.9	36
118	A Calmodulin-binding Sequence in the C-terminus of Human Cardiac Titin Kinase. FEBS Journal, 1995, 230, 752-759.	0.2	37
119	Cardiac myosin binding proteinâ€“C gene splice acceptor site mutation is associated with familial hypertrophic cardiomyopathy. Nature Genetics, 1995, 11, 438-440.	9.4	417
120	The evolution of titin and related giant muscle proteins. Journal of Molecular Evolution, 1994, 38, 395-404.	0.8	37