

Mathias Gautel

List of Publications by Year in descending order

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120
papers

17,099
citations

18436

62
h-index

19690

117
g-index

127
all docs

127
docs citations

127
times ranked

20427
citing authors

#	ARTICLE	IF	CITATIONS
1	Guidelines for the use and interpretation of assays for monitoring autophagy. <i>Autophagy</i> , 2012, 8, 445-544.	4.3	3,122
2	Reversible Unfolding of Individual Titin Immunoglobulin Domains by AFM. <i>Science</i> , 1997, 276, 1109-1112.	6.0	2,874
3	Transcriptional mechanisms regulating skeletal muscle differentiation, growth and homeostasis. <i>Nature Reviews Molecular Cell Biology</i> , 2011, 12, 349-361.	16.1	570
4	The Kinase Domain of Titin Controls Muscle Gene Expression and Protein Turnover. <i>Science</i> , 2005, 308, 1599-1603.	6.0	524
5	Cardiac myosin binding proteinâ€C gene splice acceptor site mutation is associated with familial hypertrophic cardiomyopathy. <i>Nature Genetics</i> , 1995, 11, 438-440.	9.4	417
6	Structural basis for activation of the titin kinase domain during myofibrillogenesis. <i>Nature</i> , 1998, 395, 863-869.	13.7	333
7	Mechanoenzymatics of titin kinase. <i>Proceedings of the National Academy of Sciences of the United States of America</i> , 2008, 105, 13385-13390.	3.3	311
8	The Mechanical Stability of Immunoglobulin and Fibronectin III Domains in the Muscle Protein Titin Measured by Atomic Force Microscopy. <i>Biophysical Journal</i> , 1998, 75, 3008-3014.	0.2	302
9	Obscurin, a giant sarcomeric Rho guanine nucleotide exchange factor protein involved in sarcomere assembly. <i>Journal of Cell Biology</i> , 2001, 154, 123-136.	2.3	256
10	A Molecular Map of the Interactions between Titin and Myosin-Binding Protein C. Implications for Sarcomeric Assembly in Familial Hypertrophic Cardiomyopathy. <i>FEBS Journal</i> , 1996, 235, 317-323.	0.2	249
11	The spectrin repeat: a structural platform for cytoskeletal protein assemblies. <i>FEBS Letters</i> , 2002, 513, 119-123.	1.3	249
12	Molecular Basis for Cross-Linking of Actin Filaments: Structure of the Î±-Actinin Rod. <i>Cell</i> , 1999, 98, 537-546.	13.5	237
13	Recessive mutations in EPG5 cause Vici syndrome, a multisystem disorder with defective autophagy. <i>Nature Genetics</i> , 2013, 45, 83-87.	9.4	231
14	I-Band Titin in Cardiac Muscle Is a Three-Element Molecular Spring and Is Critical for Maintaining Thin Filament Structure. <i>Journal of Cell Biology</i> , 1999, 146, 631-644.	2.3	228
15	Mutations in Î²-myosin S2 that cause familial hypertrophic cardiomyopathy (FHC) abolish the interaction with the regulatory domain of myosin-binding protein-C 1 Edited by J. Karn. <i>Journal of Molecular Biology</i> , 1999, 286, 933-949.	2.0	221
16	Congenital myopathies: disorders of excitationâ€Ccontraction coupling and muscle contraction. <i>Nature Reviews Neurology</i> , 2018, 14, 151-167.	4.9	212
17	C-terminal titin deletions cause a novel early-onset myopathy with fatal cardiomyopathy. <i>Annals of Neurology</i> , 2007, 61, 340-351.	2.8	209
18	Myosin Binding Protein C, a Phosphorylation-Dependent Force Regulator in Muscle That Controls the Attachment of Myosin Heads by Its Interaction With Myosin S2. <i>Circulation Research</i> , 2000, 86, 51-58.	2.0	200

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19	Mechanically Induced Titin Kinase Activation Studied by Force-Probe Molecular Dynamics Simulations. <i>Biophysical Journal</i> , 2005, 88, 790-804.	0.2	195
20	The sarcomeric cytoskeleton: from molecules to motion. <i>Journal of Experimental Biology</i> , 2016, 219, 135-145.	0.8	188
21	PEVK Domain of Titin: An Entropic Spring with Actin-Binding Properties. <i>Journal of Structural Biology</i> , 2002, 137, 194-205.	1.3	179
22	The Structure and Regulation of Human Muscle β -Actinin. <i>Cell</i> , 2014, 159, 1447-1460.	13.5	178
23	cAPK-phosphorylation controls the interaction of the regulatory domain of cardiac myosin binding protein C with myosin-S2 in an on-off fashion. <i>FEBS Letters</i> , 1999, 453, 254-259.	1.3	169
24	Interactions with titin and myomesin target obscurin and obscurin-like 1 to the M-band – implications for hereditary myopathies. <i>Journal of Cell Science</i> , 2008, 121, 1841-1851.	1.2	168
25	Palindromic assembly of the giant muscle protein titin in the sarcomeric Z-disk. <i>Nature</i> , 2006, 439, 229-233.	13.7	166
26	From A to Z and back? Multicompartment proteins in the sarcomere. <i>Trends in Cell Biology</i> , 2006, 16, 11-18.	3.6	163
27	The sarcomeric cytoskeleton: who picks up the strain?. <i>Current Opinion in Cell Biology</i> , 2011, 23, 39-46.	2.6	163
28	Recessive TTN truncating mutations define novel forms of core myopathy with heart disease. <i>Human Molecular Genetics</i> , 2014, 23, 980-991.	1.4	149
29	Association of the Chaperone β -crystallin with Titin in Heart Muscle. <i>Journal of Biological Chemistry</i> , 2004, 279, 7917-7924.	1.6	147
30	Two immunoglobulin-like domains of the Z-disc portion of titin interact in a conformation-dependent way with telethonin. <i>FEBS Letters</i> , 1998, 428, 111-114.	1.3	144
31	Characterization of muscle filamin isoforms suggests a possible role of β -filamin/ABP-L in sarcomeric Z-disc formation. <i>Cytoskeleton</i> , 2000, 45, 149-162.	4.4	141
32	Protein Kinase D Is a Novel Mediator of Cardiac Troponin I Phosphorylation and Regulates Myofilament Function. <i>Circulation Research</i> , 2004, 95, 1091-1099.	2.0	135
33	Transient association of titin and myosin with microtubules in nascent myofibrils directed by the MURF2 RING-finger protein. <i>Journal of Cell Science</i> , 2002, 115, 4469-4482.	1.2	131
34	A Newly Created Splice Donor Site in Exon 25 of the MyBP-C Gene Is Responsible for Inherited Hypertrophic Cardiomyopathy With Incomplete Disease Penetrance. <i>Circulation</i> , 2000, 101, 1396-1402.	1.6	114
35	Structure and Interactions of Myosin-binding Protein C Domain C0. <i>Journal of Biological Chemistry</i> , 2011, 286, 12650-12658.	1.6	114
36	Preferential skeletal muscle myosin loss in response to mechanical silencing in a novel rat intensive care unit model: underlying mechanisms. <i>Journal of Physiology</i> , 2011, 589, 2007-2026.	1.3	112

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37	Cytoskeletal protein kinases: titin and its relations in mechanosensing. Pflugers Archiv European Journal of Physiology, 2011, 462, 119-134.	1.3	111
38	Isoform Transitions of the Myosin Binding Protein C Family in Developing Human and Mouse Muscles. Circulation Research, 1998, 82, 124-129.	2.0	104
39	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
40	A molecular map of titin/connectin elasticity reveals two different mechanisms acting in series. FEBS Letters, 1996, 385, 11-14.	1.3	100
41	<i>EPG5</i>-related Vici syndrome: a paradigm of neurodevelopmental disorders with defective autophagy. Brain, 2016, 139, 765-781.	3.7	99
42	The molecular basis for sarcomere organization in vertebrate skeletal muscle. Cell, 2021, 184, 2135-2150.e13.	13.5	99
43	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
44	Distinct Sarcomeric Substrates Are Responsible for Protein Kinase D-mediated Regulation of Cardiac Myofilament Ca ²⁺ Sensitivity and Cross-bridge Cycling. Journal of Biological Chemistry, 2010, 285, 5674-5682.	1.6	96
45	The Elasticity of Single Titin Molecules Using a Two-Bead Optical Tweezers Assay. Biophysical Journal, 2004, 87, 1112-1135.	0.2	89
46	Pathogenic Mechanisms in Centronuclear Myopathies. Frontiers in Aging Neuroscience, 2014, 6, 339.	1.7	89
47	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
48	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
49	The Sarcomere and Sarcomerogenesis. Advances in Experimental Medicine and Biology, 2008, 642, 1-14.	0.8	81
50	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
51	Activation of Myocardial Contraction by the N-Terminal Domains of Myosin Binding Protein-C. Circulation Research, 2006, 98, 1290-1298.	2.0	80
52	Artifact-free high-density localization microscopy analysis. Nature Methods, 2018, 15, 689-692.	9.0	79
53	The assembly of immunoglobulin-like modules in titin: implications for muscle elasticity. Journal of Molecular Biology, 1998, 284, 761-777.	2.0	78
54	Interactions with LC3 and polyubiquitin chains link nbr1 to autophagic protein turnover. FEBS Letters, 2009, 583, 1846-1852.	1.3	78

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55	Targeted homozygous deletion of M-band titin in cardiomyocytes prevents sarcomere formation. <i>Journal of Cell Science</i> , 2006, 119, 4322-4331.	1.2	74
56	Binding of Myosin Binding Protein-C to Myosin Subfragment S2 Affects Contractility Independent of a Tether Mechanism. <i>Circulation Research</i> , 2004, 95, 930-936.	2.0	71
57	Comparing Proteins by Their Unfolding Pattern. <i>Biophysical Journal</i> , 2008, 95, 426-434.	0.2	71
58	Unfolding Forces of Titin and Fibronectin Domains Directly Measured by AFM. <i>Advances in Experimental Medicine and Biology</i> , 2000, 481, 129-141.	0.8	71
59	Calpain 1-titin interactions concentrate calpain 1 in the Z-band edges and in the N2-line region within the skeletal myofibril. <i>FEBS Journal</i> , 2005, 272, 2578-2590.	2.2	69
60	Dissecting the N-terminal Myosin Binding Site of Human Cardiac Myosin-binding Protein C. <i>Journal of Biological Chemistry</i> , 2007, 282, 9204-9215.	1.6	69
61	Structures from intact myofibrils reveal mechanism of thin filament regulation through nebulin. <i>Science</i> , 2022, 375, eabn1934.	6.0	69
62	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. <i>Circulation: Cardiovascular Genetics</i> , 2016, 9, 426-435.	5.1	67
63	The Crystal Structure of the Actin Binding Domain from $\hat{I}\pm$ -Actinin in its Closed Conformation: Structural Insight into Phospholipid Regulation of $\hat{I}\pm$ -Actinin. <i>Journal of Molecular Biology</i> , 2005, 348, 151-165.	2.0	66
64	Complete human gene structure of obscurin: implications for isoform generation by differential splicing. <i>Journal of Muscle Research and Cell Motility</i> , 2006, 26, 427-434.	0.9	65
65	Structural insight into M-band assembly and mechanics from the titin-obscurin-like-1 complex. <i>Proceedings of the National Academy of Sciences of the United States of America</i> , 2010, 107, 2908-2913.	3.3	60
66	Developmental regulation of MURF ubiquitin ligases and autophagy proteins nbr1, p62/SQSTM1 and LC3 during cardiac myofibril assembly and turnover. <i>Developmental Biology</i> , 2011, 351, 46-61.	0.9	57
67	Vici syndrome: a review. <i>Orphanet Journal of Rare Diseases</i> , 2016, 11, 21.	1.2	55
68	Developmental regulation of MURF E3 ubiquitin ligases in skeletal muscle. <i>Journal of Muscle Research and Cell Motility</i> , 2012, 33, 107-122.	0.9	46
69	Solution Scattering Suggests Cross-linking Function of Telethonin in the Complex with Titin. <i>Journal of Biological Chemistry</i> , 2003, 278, 2636-2644.	1.6	45
70	Titin Domain Patterns Correlate with the Axial Disposition of Myosin at the End of the Thick Filament. <i>Journal of Molecular Biology</i> , 1996, 259, 896-903.	2.0	44
71	Epigenetic changes as a common trigger of muscle weakness in congenital myopathies. <i>Human Molecular Genetics</i> , 2015, 24, 4636-4647.	1.4	44
72	SH3 in muscles: solution structure of the SH3 domain from nebulin. <i>Journal of Molecular Biology</i> , 1998, 276, 189-202.	2.0	40

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73	Structure, Stability and Dynamics of the Central Domain of Cardiac Myosin Binding Protein C (MyBP-C): Implications for Multidomain Assembly and Causes for Cardiomyopathy. <i>Journal of Molecular Biology</i> , 2003, 329, 745-761.	2.0	40
74	The Sarcomere and the Nucleus: Functional Links to Hypertrophy, Atrophy and Sarcopenia. <i>Advances in Experimental Medicine and Biology</i> , 2008, 642, 176-191.	0.8	40
75	The evolution of titin and related giant muscle proteins. <i>Journal of Molecular Evolution</i> , 1994, 38, 395-404.	0.8	37
76	A Calmodulin-binding Sequence in the C-terminus of Human Cardiac Titin Kinase. <i>FEBS Journal</i> , 1995, 230, 752-759.	0.2	37
77	Assembly of the cardiac I-band region of titin/connectin: expression of the cardiac-specific regions and their structural relation to the elastic segments. <i>Journal of Muscle Research and Cell Motility</i> , 1996, 17, 449-461.	0.9	36
78	Constitutive and Variable Regions of Z-disk Titin/Connectin in Myofibril Formation: A Dominant-negative Screen. <i>Cell Structure and Function</i> , 1997, 22, 95-101.	0.5	36
79	TITINdb—a computational tool to assess titin's role as a disease gene. <i>Bioinformatics</i> , 2017, 33, 3482-3485.	1.8	34
80	Making sense of missense variants in TTN-related congenital myopathies. <i>Acta Neuropathologica</i> , 2021, 141, 431-453.	3.9	34
81	Phosphoregulation of the Titin-cap Protein Telethonin in Cardiac Myocytes. <i>Journal of Biological Chemistry</i> , 2014, 289, 1282-1293.	1.6	32
82	219th ENMC International Workshop Titinopathies International database of titin mutations and phenotypes, Heemskerk, The Netherlands, 29 April–1 May 2016. <i>Neuromuscular Disorders</i> , 2017, 27, 396-407.	0.3	29
83	Current and future therapeutic approaches to the congenital myopathies. <i>Seminars in Cell and Developmental Biology</i> , 2017, 64, 191-200.	2.3	29
84	The sarcomeric cytoskeleton as a target for pharmacological intervention. <i>Current Opinion in Pharmacology</i> , 2012, 12, 347-354.	1.7	27
85	Rigid Conformation of an Immunoglobulin Domain Tandem Repeat in the A-band of the Elastic Muscle Protein Titin. <i>Journal of Molecular Biology</i> , 2007, 371, 469-480.	2.0	26
86	Evidence for a dimeric assembly of two titin/telethonin complexes induced by the telethonin C-terminus. <i>Journal of Structural Biology</i> , 2006, 155, 239-250.	1.3	25
87	Binding of Myomesin to Obscurin-Like-1 at the Muscle M-Band Provides a Strategy for Isoform-Specific Mechanical Protection. <i>Structure</i> , 2017, 25, 107-120.	1.6	25
88	A six-module human nebulin fragment bundles actin filaments and induces actin polymerization. <i>Journal of Muscle Research and Cell Motility</i> , 1998, 19, 225-235.	0.9	24
89	Binding partners of the kinase domains in <i>Drosophila</i> obscurin and their effect on the structure of the flight muscle. <i>Journal of Cell Science</i> , 2015, 128, 3386-97.	1.2	24
90	MuRF1 activity is present in cardiac mitochondria and regulates reactive oxygen species production in vivo. <i>Journal of Bioenergetics and Biomembranes</i> , 2014, 46, 173-187.	1.0	23

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91	A survey of in situ sarcomere extension in mouse skeletal muscle. <i>Journal of Muscle Research and Cell Motility</i> , 1997, 18, 465-472.	0.9	22
92	The Axial Alignment of Titin on the Muscle Thick Filament Supports Its Role as a Molecular Ruler. <i>Journal of Molecular Biology</i> , 2020, 432, 4815-4829.	2.0	21
93	A Conditional Gating Mechanism Assures the Integrity of the Molecular Force-Sensor Titin Kinase. <i>Biophysical Journal</i> , 2011, 101, 1978-1986.	0.2	20
94	The Crystal Structure of the Human Titin:Obscurin Complex Reveals a Conserved yet Specific Muscle M-Band Zipper Module. <i>Journal of Molecular Biology</i> , 2015, 427, 718-736.	2.0	20
95	Early and selective disappearance of telethonin protein from the sarcomere in neurogenic atrophy. <i>Journal of Muscle Research and Cell Motility</i> , 2001, 22, 259-264.	0.9	18
96	The spectrum of neurodevelopmental, neuromuscular and neurodegenerative disorders due to defective autophagy. <i>Autophagy</i> , 2022, 18, 496-517.	4.3	18
97	Titin ruler hypothesis not refuted. <i>Proceedings of the National Academy of Sciences of the United States of America</i> , 2015, 112, E1172-E1172.	3.3	16
98	Cardiac myosin regulatory light chain kinase modulates cardiac contractility by phosphorylating both myosin regulatory light chain and troponin I. <i>Journal of Biological Chemistry</i> , 2020, 295, 4398-4410.	1.6	16
99	When is an obscurin variant pathogenic? The impact of Arg4344Gln and Arg4444Trp variants on protein-protein interactions and protein stability. <i>Human Molecular Genetics</i> , 2021, 30, 1131-1141.	1.4	16
100	Diggin ² on U(biquitin): A Novel Method for the Identification of Physiological E3 Ubiquitin Ligase Substrates. <i>Cell Biochemistry and Biophysics</i> , 2013, 67, 127-138.	0.9	15
101	Order from disorder in the sarcomere: FATZ forms a fuzzy but tight complex and phase-separated condensates with β -actinin. <i>Science Advances</i> , 2021, 7, .	4.7	15
102	Increasing evidence of mechanical force as a functional regulator in smooth muscle myosin light chain kinase. <i>ELife</i> , 2017, 6, .	2.8	15
103	Sub-diffraction error mapping for localisation microscopy images. <i>Nature Communications</i> , 2021, 12, 5611.	5.8	14
104	Clinical utility gene card for: Vici Syndrome. <i>European Journal of Human Genetics</i> , 2014, 22, 435-435.	1.4	13
105	Reply: Hereditary myopathy with early respiratory failure is caused by mutations in the titin FN3 119 domain. <i>Brain</i> , 2014, 137, e279-e279.	3.7	13
106	Myopalladin knockout mice develop cardiac dilation and show a maladaptive response to mechanical pressure overload. <i>ELife</i> , 2021, 10, .	2.8	12
107	Kinase recognition by calmodulin: modeling the interaction with the autoinhibitory region of human cardiac titin kinase ¹¹ Edited by J. Thornton. <i>Journal of Molecular Biology</i> , 2001, 306, 81-95.	2.0	8
108	Myofibrillar tightly bound calcium in skeletal muscle fibers: a possible role of this cation in titin strands aggregation. <i>FEBS Letters</i> , 2004, 556, 271-275.	1.3	8

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109	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
110	Autopsy findings in <i>EPG5</i> -related Vici syndrome with antenatal onset. American Journal of Medical Genetics, Part A, 2017, 173, 2522-2527.	0.7	6
111	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
112	Molecular noise filtering in the β^2 -adrenergic signaling network by phospholamban pentamers. Cell Reports, 2021, 36, 109448.	2.9	5
113	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
114	Gett'N-WASP Stripes. Science, 2010, 330, 1491-1492.	6.0	4
115	Reply: Aberrant splicing induced by the most common <i>EPG5</i> mutation in an individual with Vici syndrome. Brain, 2016, 139, e53-e53.	3.7	4
116	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
117	The molecular basis for sarcomere organization in vertebrate skeletal muscle. Microscopy and Microanalysis, 2021, 27, 2832-2835.	0.2	1
118	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
119	Editorial June 2010. Journal of Muscle Research and Cell Motility, 2010, 31, 1-1.	0.9	0
120	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