

# Mathias Gautel

## List of Publications by Year in descending order

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120  
papers

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citations

18482  
62  
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19749  
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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.	9.1	3,122
2	Reversible Unfolding of Individual Titin Immunoglobulin Domains by AFM. <i>Science</i> , 1997, 276, 1109-1112.	12.6	2,874
3	Transcriptional mechanisms regulating skeletal muscle differentiation, growth and homeostasis. <i>Nature Reviews Molecular Cell Biology</i> , 2011, 12, 349-361.	37.0	570
4	The Kinase Domain of Titin Controls Muscle Gene Expression and Protein Turnover. <i>Science</i> , 2005, 308, 1599-1603.	12.6	524
5	Cardiac myosin binding protein-C gene splice acceptor site mutation is associated with familial hypertrophic cardiomyopathy. <i>Nature Genetics</i> , 1995, 11, 438-440.	21.4	417
6	Structural basis for activation of the titin kinase domain during myofibrillogenesis. <i>Nature</i> , 1998, 395, 863-869.	27.8	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.	7.1	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.	0.5	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.	5.2	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.	2.8	249
12	Molecular Basis for Cross-Linking of Actin Filaments: Structure of the $\alpha$ -Actinin Rod. <i>Cell</i> , 1999, 98, 537-546.	28.9	237
13	Recessive mutations in EPG5 cause Vici syndrome, a multisystem disorder with defective autophagy. <i>Nature Genetics</i> , 2013, 45, 83-87.	21.4	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.	5.2	228
15	Mutations in $\beta$ -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.	4.2	221
16	Congenital myopathies: disorders of excitation-contraction coupling and muscle contraction. <i>Nature Reviews Neurology</i> , 2018, 14, 151-167.	10.1	212
17	C-terminal titin deletions cause a novel early-onset myopathy with fatal cardiomyopathy. <i>Annals of Neurology</i> , 2007, 61, 340-351.	5.3	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.	4.5	200

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19	Mechanically Induced Titin Kinase Activation Studied by Force-Probe Molecular Dynamics Simulations. Biophysical Journal, 2005, 88, 790-804.	0.5	195
20	The sarcomeric cytoskeleton: from molecules to motion. Journal of Experimental Biology, 2016, 219, 135-145.	1.7	188
21	PEVK Domain of Titin: An Entropic Spring with Actin-Binding Properties. Journal of Structural Biology, 2002, 137, 194-205.	2.8	179
22	The Structure and Regulation of Human Muscle $\beta$ -Actinin. Cell, 2014, 159, 1447-1460.	28.9	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. FEBS Letters, 1999, 453, 254-259.	2.8	169
24	Interactions with titin and myomesin target obscurin and obscurin-like 1 to the M-band – implications for hereditary myopathies. Journal of Cell Science, 2008, 121, 1841-1851.	2.0	168
25	Palindromic assembly of the giant muscle protein titin in the sarcomeric Z-disk. Nature, 2006, 439, 229-233.	27.8	166
26	From A to Z and back? Multicompartment proteins in the sarcomere. Trends in Cell Biology, 2006, 16, 11-18.	7.9	163
27	The sarcomeric cytoskeleton: who picks up the strain?. Current Opinion in Cell Biology, 2011, 23, 39-46.	5.4	163
28	Recessive TTN truncating mutations define novel forms of core myopathy with heart disease. Human Molecular Genetics, 2014, 23, 980-991.	2.9	149
29	Association of the Chaperone $\beta$ -crystallin with Titin in Heart Muscle. Journal of Biological Chemistry, 2004, 279, 7917-7924.	3.4	147
30	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.	2.8	144
31	Characterization of muscle filamin isoforms suggests a possible role of $\gamma$ -filamin/ABP-L in sarcomeric Z-disc formation. Cytoskeleton, 2000, 45, 149-162.	4.4	141
32	Protein Kinase D Is a Novel Mediator of Cardiac Troponin I Phosphorylation and Regulates Myofilament Function. Circulation Research, 2004, 95, 1091-1099.	4.5	135
33	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.	2.0	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. Circulation, 2000, 101, 1396-1402.	1.6	114
35	Structure and Interactions of Myosin-binding Protein C Domain C0. Journal of Biological Chemistry, 2011, 286, 12650-12658.	3.4	114
36	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.	2.9	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.	2.8	111
38	Isoform Transitions of the Myosin Binding Protein C Family in Developing Human and Mouse Muscles. Circulation Research, 1998, 82, 124-129.	4.5	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.	7.1	103
40	A molecular map of titin/connectin elasticity reveals two different mechanisms acting in series. FEBS Letters, 1996, 385, 11-14.	2.8	100
41	<i>EPG5</i>-related Vici syndrome: a paradigm of neurodevelopmental disorders with defective autophagy. Brain, 2016, 139, 765-781.	7.6	99
42	The molecular basis for sarcomere organization in vertebrate skeletal muscle. Cell, 2021, 184, 2135-2150.e13.	28.9	99
43	Crystal structures of human cardiac $\beta$ -myosin II S2- $\beta$ 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.	7.1	97
44	Distinct Sarcomeric Substrates Are Responsible for Protein Kinase D-mediated Regulation of Cardiac Myofilament Ca <sup>2+</sup> Sensitivity and Cross-bridge Cycling. Journal of Biological Chemistry, 2010, 285, 5674-5682.	3.4	96
45	The Elasticity of Single Titin Molecules Using a Two-Bead Optical Tweezers Assay. Biophysical Journal, 2004, 87, 1112-1135.	0.5	89
46	Pathogenic Mechanisms in Centronuclear Myopathies. Frontiers in Aging Neuroscience, 2014, 6, 339.	3.4	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.	4.2	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.	2.0	85
49	The Sarcomere and Sarcomerogenesis. Advances in Experimental Medicine and Biology, 2008, 642, 1-14.	1.6	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.	3.3	80
51	Activation of Myocardial Contraction by the N-Terminal Domains of Myosin Binding Protein-C. Circulation Research, 2006, 98, 1290-1298.	4.5	80
52	Artifact-free high-density localization microscopy analysis. Nature Methods, 2018, 15, 689-692.	19.0	79
53	The assembly of immunoglobulin-like modules in titin: implications for muscle elasticity. Journal of Molecular Biology, 1998, 284, 761-777.	4.2	78
54	Interactions with LC3 and polyubiquitin chains link nbr1 to autophagic protein turnover. FEBS Letters, 2009, 583, 1846-1852.	2.8	78

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55	Targeted homozygous deletion of M-band titin in cardiomyocytes prevents sarcomere formation. Journal of Cell Science, 2006, 119, 4322-4331.	2.0	74
56	Binding of Myosin Binding Protein-C to Myosin Subfragment S2 Affects Contractility Independent of a Tether Mechanism. Circulation Research, 2004, 95, 930-936.	4.5	71
57	Comparing Proteins by Their Unfolding Pattern. Biophysical Journal, 2008, 95, 426-434.	0.5	71
58	Unfolding Forces of Titin and Fibronectin Domains Directly Measured by AFM. Advances in Experimental Medicine and Biology, 2000, 481, 129-141.	1.6	71
59	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.	4.7	69
60	Dissecting the N-terminal Myosin Binding Site of Human Cardiac Myosin-binding Protein C. Journal of Biological Chemistry, 2007, 282, 9204-9215.	3.4	69
61	Structures from intact myofibrils reveal mechanism of thin filament regulation through nebulin. Science, 2022, 375, eabn1934.	12.6	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. Circulation: Cardiovascular Genetics, 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. Journal of Molecular Biology, 2005, 348, 151-165.	4.2	66
64	Complete human gene structure of obscurin: implications for isoform generation by differential splicing. Journal of Muscle Research and Cell Motility, 2006, 26, 427-434.	2.0	65
65	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.	7.1	60
66	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.	2.0	57
67	Vici syndrome: a review. Orphanet Journal of Rare Diseases, 2016, 11, 21.	2.7	55
68	Developmental regulation of MURF E3 ubiquitin ligases in skeletal muscle. Journal of Muscle Research and Cell Motility, 2012, 33, 107-122.	2.0	46
69	Solution Scattering Suggests Cross-linking Function of Telethonin in the Complex with Titin. Journal of Biological Chemistry, 2003, 278, 2636-2644.	3.4	45
70	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.	4.2	44
71	Epigenetic changes as a common trigger of muscle weakness in congenital myopathies. Human Molecular Genetics, 2015, 24, 4636-4647.	2.9	44
72	SH3 in muscles: solution structure of the SH3 domain from nebulin. Journal of Molecular Biology, 1998, 276, 189-202.	4.2	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. Journal of Molecular Biology, 2003, 329, 745-761.	4.2	40
74	The Sarcomere and the Nucleus: Functional Links to Hypertrophy, Atrophy and Sarcopenia. Advances in Experimental Medicine and Biology, 2008, 642, 176-191.	1.6	40
75	The evolution of titin and related giant muscle proteins. Journal of Molecular Evolution, 1994, 38, 395-404.	1.8	37
76	A Calmodulin-binding Sequence in the C-terminus of Human Cardiac Titin Kinase. FEBS Journal, 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. Journal of Muscle Research and Cell Motility, 1996, 17, 449-461.	2.0	36
78	Constitutive and Variable Regions of Z-disk Titin/Connectin in Myofibril Formation: A Dominant-negative Screen.. Cell Structure and Function, 1997, 22, 95-101.	1.1	36
79	TITINdb—a computational tool to assess titin’s role as a disease gene. Bioinformatics, 2017, 33, 3482-3485.	4.1	34
80	Making sense of missense variants in TTN-related congenital myopathies. Acta Neuropathologica, 2021, 141, 431-453.	7.7	34
81	Phosphoregulation of the Titin-cap Protein Telethonin in Cardiac Myocytes. Journal of Biological Chemistry, 2014, 289, 1282-1293.	3.4	32
82	219th ENMC International Workshop Titinopathies International database of titin mutations and phenotypes, Heemskerk, The Netherlands, 29 April–1 May 2016. Neuromuscular Disorders, 2017, 27, 396-407.	0.6	29
83	Current and future therapeutic approaches to the congenital myopathies. Seminars in Cell and Developmental Biology, 2017, 64, 191-200.	5.0	29
84	The sarcomeric cytoskeleton as a target for pharmacological intervention. Current Opinion in Pharmacology, 2012, 12, 347-354.	3.5	27
85	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.	4.2	26
86	Evidence for a dimeric assembly of two titin/telethonin complexes induced by the telethonin C-terminus. Journal of Structural Biology, 2006, 155, 239-250.	2.8	25
87	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.	3.3	25
88	A six-module human nebulin fragment bundles actin filaments and induces actin polymerization. Journal of Muscle Research and Cell Motility, 1998, 19, 225-235.	2.0	24
89	Binding partners of the kinase domains in <i>Drosophila</i> obscurin and their effect on the structure of the flight muscle. Journal of Cell Science, 2015, 128, 3386-97.	2.0	24
90	MuRF1 activity is present in cardiac mitochondria and regulates reactive oxygen species production in vivo. Journal of Bioenergetics and Biomembranes, 2014, 46, 173-187.	2.3	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.	2.0	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.	4.2	21
93	A Conditional Gating Mechanism Assures the Integrity of the Molecular Force-Sensor Titin Kinase. <i>Biophysical Journal</i> , 2011, 101, 1978-1986.	0.5	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.	4.2	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.	2.0	18
96	The spectrum of neurodevelopmental, neuromuscular and neurodegenerative disorders due to defective autophagy. <i>Autophagy</i> , 2022, 18, 496-517.	9.1	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.	7.1	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.	3.4	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.	2.9	16
100	Diggin' 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.	1.8	15
101	Order from disorder in the sarcomere: FATZ forms a fuzzy but tight complex and phase-separated condensates with $\beta$ -actinin. <i>Science Advances</i> , 2021, 7, .	10.3	15
102	Increasing evidence of mechanical force as a functional regulator in smooth muscle myosin light chain kinase. <i>ELife</i> , 2017, 6, .	6.0	15
103	Sub-diffraction error mapping for localisation microscopy images. <i>Nature Communications</i> , 2021, 12, 5611.	12.8	14
104	Clinical utility gene card for: Vici Syndrome. <i>European Journal of Human Genetics</i> , 2014, 22, 435-435.	2.8	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.	7.6	13
106	Myopalladin knockout mice develop cardiac dilation and show a maladaptive response to mechanical pressure overload. <i>ELife</i> , 2021, 10, .	6.0	12
107	Kinase recognition by calmodulin: modeling the interaction with the autoinhibitory region of human cardiac titin kinase <sup>11</sup> Edited by J. Thornton. <i>Journal of Molecular Biology</i> , 2001, 306, 81-95.	4.2	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.	2.8	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.	3.4	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.	1.2	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.	2.8	5
112	Molecular noise filtering in the $\beta^2$ -adrenergic signaling network by phospholamban pentamers. Cell Reports, 2021, 36, 109448.	6.4	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.	2.8	4
114	Gett'N-WASP Stripes. Science, 2010, 330, 1491-1492.	12.6	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.	7.6	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.	0.8	1
117	The molecular basis for sarcomere organization in vertebrate skeletal muscle. Microscopy and Microanalysis, 2021, 27, 2832-2835.	0.4	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.	2.8	1
119	Editorial June 2010. Journal of Muscle Research and Cell Motility, 2010, 31, 1-1.	2.0	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.	2.0	0