Sébastien Fribourg

List of Publications by Year in descending order

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#	Article	IF	CITATIONS
1	Mutations in the XPD helicase gene result in XP and TTD phenotypes, preventing interaction between XPD and the p44 subunit of TFIIH. Nature Genetics, 1998, 20, 184-188.	21.4	320
2	Mutations of POLR3A Encoding a Catalytic Subunit of RNA Polymerase Pol III Cause a Recessive Hypomyelinating Leukodystrophy. American Journal of Human Genetics, 2011, 89, 415-423.	6.2	219
3	Structural Basis for the Recognition of a Nucleoporin FG Repeat by the NTF2-like Domain of the TAP/p15 mRNA Nuclear Export Factor. Molecular Cell, 2001, 8, 645-656.	9.7	211
4	Impaired ribosome biogenesis in Diamond-Blackfan anemia. Blood, 2007, 109, 1275-1283.	1.4	202
5	The archaeal exosome core is a hexameric ring structure with three catalytic subunits. Nature Structural and Molecular Biology, 2005, 12, 575-581.	8.2	198
6	Molecular Structure of Human TFIIH. Cell, 2000, 102, 599-607.	28.9	175
7	Clinical spectrum of 4H leukodystrophy caused by <i>POLR3A</i> and <i>POLR3B</i> mutations. Neurology, 2014, 83, 1898-1905.	1.1	170
8	A novel mode of RBD-protein recognition in the Y14–Mago complex. Nature Structural and Molecular Biology, 2003, 10, 433-439.	8.2	150
9	Recessive Mutations in POLR3B, Encoding the Second Largest Subunit of Pol III, Cause a Rare Hypomyelinating Leukodystrophy. American Journal of Human Genetics, 2011, 89, 652-655.	6.2	139
10	Recessive mutations in POLR1C cause a leukodystrophy by impairing biogenesis of RNA polymerase III. Nature Communications, 2015, 6, 7623.	12.8	127
11	Inborn errors in RNA polymerase III underlie severe varicella zoster virus infections. Journal of Clinical Investigation, 2017, 127, 3543-3556.	8.2	125
12	Deciphering correct strategies for multiprotein complex assembly by co-expression: Application to complexes as large as the histone octamer. Journal of Structural Biology, 2011, 175, 178-188.	2.8	116
13	Mutation of ribosomal protein RPS24 in Diamond-Blackfan anemia results in a ribosome biogenesis disorder. Human Molecular Genetics, 2008, 17, 1253-1263.	2.9	100
14	RPS19 mutations in patients with Diamond-Blackfan anemia. Human Mutation, 2008, 29, 911-920.	2.5	94
15	Peptides derived from the bifunctional kinase/RNase enzyme IRE1α modulate IRE1α activity and protect cells from endoplasmic reticulum stress. FASEB Journal, 2011, 25, 3115-3129.	0.5	71
16	Dissecting the interaction network of multiprotein complexes by pairwise coexpression of subunits in E. coli11Edited by K. Nagai. Journal of Molecular Biology, 2001, 306, 363-373.	4.2	64
17	Molecular basis of Diamond Blackfan anemia: structure and function analysis of RPS19. Nucleic Acids Research, 2007, 35, 5913-5921.	14.5	56
18	Exploring TAR–RNA aptamer loop–loop interaction by X-ray crystallography, UV spectroscopy and surface plasmon resonance. Nucleic Acids Research, 2008, 36, 7146-7156.	14.5	54

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19	Varicella-zoster virus CNS vasculitis and RNA polymerase III gene mutation in identical twins. Neurology: Neuroimmunology and NeuroInflammation, 2018, 5, e500.	6.0	49
20	Structural similarity in the absence of sequence homology of the messenger RNA export factors Mtr2 and p15. EMBO Reports, 2003, 4, 699-703.	4.5	48
21	Structure-function analysis of hRPC62 provides insights into RNA polymerase III transcription initiation. Nature Structural and Molecular Biology, 2011, 18, 352-358.	8.2	43
22	The structure of the CstF-77 homodimer provides insights into CstF assembly. Nucleic Acids Research, 2007, 35, 4515-4522.	14.5	42
23	Structural and functional aspects of winged-helix domains at the core of transcription initiation complexes. Transcription, 2012, 3, 2-7.	3.1	42
24	Clinical spectrum of POLR3-related leukodystrophy caused by biallelic <i>POLR1C</i> pathogenic variants. Neurology: Genetics, 2019, 5, e369.	1.9	38
25	An essential role for Clp1 in assembly of polyadenylation complex CF IA and Pol II transcription termination. Nucleic Acids Research, 2012, 40, 1226-1239.	14.5	31
26	Locked Tether Formation by Cooperative Folding of Rna14p Monkeytail and Rna15p Hinge Domains in the Yeast CF IA Complex. Structure, 2011, 19, 534-545.	3.3	29
27	Structural Characterization of the Cysteine-rich Domain of TFIIH p44 Subunit. Journal of Biological Chemistry, 2000, 275, 31963-31971.	3.4	28
28	Solution Structure of the C-terminal Domain of TFIIH P44 Subunit Reveals a Novel Type of C4C4 Ring Domain Involved in Protein-Protein Interactions. Journal of Biological Chemistry, 2005, 280, 20785-20792.	3.4	28
29	Crucial role of the Rcl1p–Bms1p interaction for yeast pre-ribosomal RNA processing. Nucleic Acids Research, 2014, 42, 10161-10172.	14.5	26
30	Mutations in the amino-terminal domain of the human poly(ADP-ribose) polymerase that affect its catalytic activity but not its DNA binding capacity. FEBS Letters, 1996, 399, 313-316.	2.8	25
31	The Npa1p complex chaperones the assembly of the earliest eukaryotic large ribosomal subunit precursor. PLoS Genetics, 2018, 14, e1007597.	3.5	23
32	De novo variants in POLR3B cause ataxia, spasticity, and demyelinating neuropathy. American Journal of Human Genetics, 2021, 108, 186-193.	6.2	19
33	Hexameric architecture of CstF supported by CstF-50 homodimerization domain structure. Rna, 2011, 17, 412-418.	3.5	17
34	Structural insights into the 3′-end mRNA maturation machinery: Snapshot on polyadenylation signal recognition. Biochimie, 2019, 164, 105-110.	2.6	17
35	Expression of FLAG Fusion Proteins in Insect Cells: Application to the Multi-subunit Transcription/DNA Repair Factor TFIIH. Protein Expression and Purification, 2002, 24, 513-523.	1.3	15
36	Structural analysis of human RPC32β–RPC62 complex. Journal of Structural Biology, 2015, 192, 313-319.	2.8	11

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37	Distinct roles of Pcf11 zinc-binding domains in pre-mRNA 3′-end processing. Nucleic Acids Research, 2017, 45, 10115-10131.	14.5	11
38	Pwp2 mediates UTP-B assembly via two structurally independent domains. Scientific Reports, 2017, 7, 3169.	3.3	9
39	The hRPC62 subunit of human RNA polymerase III displays helicase activity. Nucleic Acids Research, 2019, 47, 10313-10326.	14.5	9
40	Domain definition and interaction mapping for the endonuclease complex hNob1/hPno1. RNA Biology, 2018, 15, 1174-1180.	3.1	8
41	In vitro dimerization of human RIO2 kinase. RNA Biology, 2019, 16, 1633-1642.	3.1	7
42	Structural basis for ATP loss by Clp1p in a G135R mutant protein. Biochimie, 2014, 101, 203-207.	2.6	5
43	Structural characterization of the yeast CF IA complex through a combination of mass spectrometry approaches. International Journal of Mass Spectrometry, 2017, 420, 57-66.	1.5	5
44	Crystal structure of SFPQ-NONO heterodimer. Biochimie, 2022, 198, 1-7.	2.6	5
45	Chemical shift assignments of a new folded domain from yeast Pcf11. Biomolecular NMR Assignments, 2015, 9, 421-425.	0.8	4
46	Sqt1p is an eight-bladed WD40 protein. Acta Crystallographica Section F, Structural Biology Communications, 2016, 72, 59-64.	0.8	2
47	Mutations of POLR3A Encoding a Catalytic Subunit of RNA Polymerase Pol III Cause a Recessive Hypomyelinating Leukodystrophy p415. American Journal of Human Genetics, 2012, 91, 972.	6.2	1
48	Structural and interaction analysis of the Rrp5 Câ€ŧerminal region. FEBS Open Bio, 2018, 8, 1605-1614.	2.3	1