Sarah J Tabrizi

List of Publications by Citations

141 papers	9,229 citations	48 h-index	95 g-index
179 ext. papers	11,256 ext. citations	9.1 avg, IF	5.93 L-index

#	Paper	IF	Citations
141	Biological and clinical manifestations of Huntington's disease in the longitudinal TRACK-HD study: cross-sectional analysis of baseline data. *Lancet Neurology, The*, **2009**, 8, 791-801	24.1	721
140	Huntington disease. *Nature Reviews Disease Primers*, **2015**, 1, 15005	51.1	672
139	Huntington disease: natural history, biomarkers and prospects for therapeutics. *Nature Reviews Neurology*, **2014**, 10, 204-16	15	600
138	Predictors of phenotypic progression and disease onset in premanifest and early-stage Huntington's disease in the TRACK-HD study: analysis of 36-month observational data. *Lancet Neurology, The*, **2013**, 12, 637-49	24.1	557
137	Biological and clinical changes in premanifest and early stage Huntington's disease in the TRACK-HD study: the 12-month longitudinal analysis. *Lancet Neurology, The*, **2011**, 10, 31-42	24.1	443
136	A novel pathogenic pathway of immune activation detectable before clinical onset in Huntington's disease. *Journal of Experimental Medicine*, **2008**, 205, 1869-77	16.6	437
135	Potential endpoints for clinical trials in premanifest and early Huntington's disease in the TRACK-HD study: analysis of 24 month observational data. *Lancet Neurology, The*, **2012**, 11, 42-53	24.1	392
134	Microglial activation in presymptomatic Huntington's disease gene carriers. *Brain*, **2007**, 130, 1759-66	11.2	324
133	Targeting Huntingtin Expression in Patients with Huntington's Disease. *New England Journal of Medicine*, **2019**, 380, 2307-2316	59.2	319
132	Therapies targeting DNA and RNA in Huntington's disease. *Lancet Neurology, The*, **2017**, 16, 837-847	24.1	175
131	Proteomic profiling of plasma in Huntington's disease reveals neuroinflammatory activation and biomarker candidates. *Journal of Proteome Research*, **2007**, 6, 2833-40	5.6	173
130	Neurofilament light protein in blood as a potential biomarker of neurodegeneration in Huntington's disease: a retrospective cohort analysis. *Lancet Neurology, The*, **2017**, 16, 601-609	24.1	172
129	Identification of genetic variants associated with Huntington's disease progression: a genome-wide association study. *Lancet Neurology, The*, **2017**, 16, 701-711	24.1	161
128	C9orf72 expansions are the most common genetic cause of Huntington disease phenocopies. *Neurology*, **2014**, 82, 292-9	6.5	152
127	Quantification of mutant huntingtin protein in cerebrospinal fluid from Huntington's disease patients. *Journal of Clinical Investigation*, **2015**, 125, 1979-86	15.9	144
126	White matter connections reflect changes in voluntary-guided saccades in pre-symptomatic Huntington's disease. *Brain*, **2008**, 131, 196-204	11.2	143
125	DNA repair pathways underlie a common genetic mechanism modulating onset in polyglutamine diseases. *Annals of Neurology*, **2016**, 79, 983-90	9.4	135

124	Prevalence of adult Huntington's disease in the UK based on diagnoses recorded in general practice records. *Journal of Neurology, Neurosurgery and Psychiatry*, **2013**, 84, 1156-60	5.5	130
123	An event-based model for disease progression and its application in familial Alzheimer's disease and Huntington's disease. *NeuroImage*, **2012**, 60, 1880-9	7.9	125
122	HTT-lowering reverses Huntington's disease immune dysfunction caused by NFB pathway dysregulation. *Brain*, **2014**, 137, 819-33	11.2	109
121	Mutant huntingtin impairs immune cell migration in Huntington disease. *Journal of Clinical Investigation*, **2012**, 122, 4737-47	15.9	105
120	Huntingtin Lowering Strategies for Disease Modification in Huntington's Disease. *Neuron*, **2019**, 101, 801-819	13.9	102
119	An exploratory double-blind, randomized clinical trial with selisistat, a SirT1 inhibitor, in patients with Huntington's disease. *British Journal of Clinical Pharmacology*, **2015**, 79, 465-76	3.8	98
118	Early atrophy of pallidum and accumbens nucleus in Huntington's disease. *Journal of Neurology*, **2011**, 258, 412-20	5.5	98
117	Evaluation of longitudinal 12 and 24 month cognitive outcomes in premanifest and early Huntington's disease. *Journal of Neurology, Neurosurgery and Psychiatry*, **2012**, 83, 687-94	5.5	97
116	Mutant huntingtin fragmentation in immune cells tracks Huntington's disease progression. *Journal of Clinical Investigation*, **2012**, 122, 3731-6	15.9	97
115	Huntington's disease phenocopies are clinically and genetically heterogeneous. *Movement Disorders*, **2008**, 23, 716-20	7	93
114	Cannabinoid receptor 2 signaling in peripheral immune cells modulates disease onset and severity in mouse models of Huntington's disease. *Journal of Neuroscience*, **2012**, 32, 18259-68	6.6	92
113	Compensation in Preclinical Huntington's Disease: Evidence From the Track-On HD Study. *EBioMedicine*, **2015**, 2, 1420-9	8.8	91
112	The progression of regional atrophy in premanifest and early Huntington's disease: a longitudinal voxel-based morphometry study. *Journal of Neurology, Neurosurgery and Psychiatry*, **2010**, 81, 756-63	5.5	90
111	The pathogenic exon 1 HTT protein is produced by incomplete splicing in Huntington's disease patients. *Scientific Reports*, **2017**, 7, 1307	4.9	89
110	Functional compensation of motor function in pre-symptomatic Huntington's disease. *Brain*, **2009**, 132, 1624-32	11.2	87
109	Huntington disease: new insights into molecular pathogenesis and therapeutic opportunities. *Nature Reviews Neurology*, **2020**, 16, 529-546	15	80
108	Clinical impairment in premanifest and early Huntington's disease is associated with regionally specific atrophy. *Human Brain Mapping*, **2013**, 34, 519-29	5.9	77
107	Abnormal peripheral chemokine profile in Huntington's disease. *PLOS Currents*, **2011**, 3, RRN1231		73

106	A SNP in the HTT promoter alters NF-κB binding and is a bidirectional genetic modifier of Huntington disease. *Nature Neuroscience*, **2015**, 18, 807-16	25.5	70
105	Structural MRI in Huntington's disease and recommendations for its potential use in clinical trials. *Neuroscience and Biobehavioral Reviews*, **2013**, 37, 480-90	9	70
104	Selective vulnerability of Rich Club brain regions is an organizational principle of structural connectivity loss in Huntington's disease. *Brain*, **2015**, 138, 3327-44	11.2	66
103	Observing Huntington's Disease: the European Huntington's Disease Network's REGISTRY. *PLOS Currents*, **2010**, 2,		64
102	A genetic association study of glutamine-encoding DNA sequence structures, somatic CAG expansion, and DNA repair gene variants, with Huntington disease clinical outcomes. *EBioMedicine*, **2019**, 48, 568-580	8.8	63
101	Huntington's disease phenocopy syndromes. *Current Opinion in Neurology*, **2007**, 20, 681-7	7.1	63
100	MSH3 modifies somatic instability and disease severity in Huntington's and myotonic dystrophy type 1. *Brain*, **2019**,	11.2	57
99	Motor, cognitive, and functional declines contribute to a single progressive factor in early HD. *Neurology*, **2017**, 89, 2495-2502	6.5	57
98	Bone marrow transplantation confers modest benefits in mouse models of Huntington's disease. *Journal of Neuroscience*, **2012**, 32, 133-42	6.6	57
97	Biological and clinical characteristics of gene carriers far from predicted onset in the Huntington's disease Young Adult Study (HD-YAS): a cross-sectional analysis. *Lancet Neurology, The*, **2020**, 19, 502-512	24.1	56
96	FAN1 modifies Huntington's disease progression by stabilizing the expanded HTT CAG repeat. *Human Molecular Genetics*, **2019**, 28, 650-661	5.6	56
95	Brain Regions Showing White Matter Loss in Huntington's Disease Are Enriched for Synaptic and Metabolic Genes. *Biological Psychiatry*, **2018**, 83, 456-465	7.9	54
94	Clinical Features of Huntington's Disease. *Advances in Experimental Medicine and Biology*, **2018**, 1049, 1-28	3.6	53
93	White matter integrity in premanifest and early Huntington's disease is related to caudate loss and disease progression. *Cortex*, **2014**, 52, 98-112	3.8	46
92	The impact of occipital lobe cortical thickness on cognitive task performance: An investigation in Huntington's Disease. *Neuropsychologia*, **2015**, 79, 138-46	3.2	42
91	Neurofilament light protein in blood predicts regional atrophy in Huntington disease. *Neurology*, **2018**, 90, e717-e723	6.5	42
90	Automated quantification of caudate atrophy by local registration of serial MRI: evaluation and application in Huntington's disease. *NeuroImage*, **2009**, 47, 1659-65	7.9	38
89	Cerebrospinal fluid total tau concentration predicts clinical phenotype in Huntington's disease. *Journal of Neurochemistry*, **2016**, 139, 22-5	6	37

88	Huntington disease. *Handbook of Clinical Neurology / Edited By P J Vinken and G W Bruyn*, **2018**, 147, 255-278	3	37
87	Characterisation of immune cell function in fragment and full-length Huntington's disease mouse models. *Neurobiology of Disease*, **2015**, 73, 388-98	7.5	37
86	Cerebrospinal Fluid Inflammatory Biomarkers Reflect Clinical Severity in Huntington's Disease. *PLoS ONE*, **2016**, 11, e0163479	3.7	35
85	Mouse models for neurological disease. *Lancet Neurology, The*, **2002**, 1, 215-24	24.1	34
84	RNA-Seq of Huntington's disease patient myeloid cells reveals innate transcriptional dysregulation associated with proinflammatory pathway activation. *Human Molecular Genetics*, **2016**, 25, 2893-2904	5.6	33
83	Huntington's disease blood and brain show a common gene expression pattern and share an immune signature with Alzheimer's disease. *Scientific Reports*, **2017**, 7, 44849	4.9	31
82	An image-based model of brain volume biomarker changes in Huntington's disease. *Annals of Clinical and Translational Neurology*, **2018**, 5, 570-582	5.3	31
81	Movement Disorder Society Task Force Viewpoint: Huntington's Disease Diagnostic Categories. *Movement Disorders Clinical Practice*, **2019**, 6, 541-546	2.2	30
80	In vivo characterization of white matter pathology in premanifest huntington's disease. *Annals of Neurology*, **2018**, 84, 497-504	9.4	29
79	Evaluation of multi-modal, multi-site neuroimaging measures in Huntington's disease: Baseline results from the PADDINGTON study. *NeuroImage: Clinical*, **2012**, 2, 204-11	5.3	29
78	Topological length of white matter connections predicts their rate of atrophy in premanifest Huntington's disease. *JCI Insight*, **2017**, 2,	9.9	27
77	Basal ganglia-cortical structural connectivity in Huntington's disease. *Human Brain Mapping*, **2015**, 36, 1728-40	5.9	26
76	Stimulating neural plasticity with real-time fMRI neurofeedback in Huntington's disease: A proof of concept study. *Human Brain Mapping*, **2018**, 39, 1339-1353	5.9	24
75	Mutant huntingtin and neurofilament light have distinct longitudinal dynamics in Huntington's disease. *Science Translational Medicine*, **2020**, 12,	17.5	24
74	Overlap between age-at-onset and disease-progression determinants in Huntington disease. *Neurology*, **2018**, 90, e2099-e2106	6.5	22
73	Association of CAG Repeats With Long-term Progression in Huntington Disease. *JAMA Neurology*, **2019**, 76, 1375-1385	17.2	22
72	White matter predicts functional connectivity in premanifest Huntington's disease. *Annals of Clinical and Translational Neurology*, **2017**, 4, 106-118	5.3	21
71	The human motor cortex microcircuit: insights for neurodegenerative disease. *Nature Reviews Neuroscience*, **2020**, 21, 401-415	13.5	20

70	A critical evaluation of inflammatory markers in Huntington's Disease plasma. *Journal of Huntingtons Disease*, **2013**, 2, 125-34	1.9	19
69	Longitudinal changes in functional connectivity of cortico-basal ganglia networks in manifests and premanifest huntington's disease. *Human Brain Mapping*, **2016**, 37, 4112-4128	5.9	18
68	Structural and functional brain network correlates of depressive symptoms in premanifest Huntington's disease. *Human Brain Mapping*, **2017**, 38, 2819-2829	5.9	17
67	Robust Markers and Sample Sizes for Multicenter Trials of Huntington Disease. *Annals of Neurology*, **2020**, 87, 751-762	9.4	14
66	Structural imaging in premanifest and manifest Huntington disease. *Handbook of Clinical Neurology / Edited By P J Vinken and G W Bruyn*, **2017**, 144, 247-261	3	14
65	Rate and acceleration of whole-brain atrophy in premanifest and early Huntington's disease. *Movement Disorders*, **2010**, 25, 888-95	7	12
64	Myostatin inhibition prevents skeletal muscle pathophysiology in Huntington's disease mice. *Scientific Reports*, **2017**, 7, 14275	4.9	11
63	A17 HD brain-train: neuroplasticity as a target to improve function in huntingtons disease. *Journal of Neurology, Neurosurgery and Psychiatry*, **2016**, 87, A5.3-A5	5.5	10
62	Dynamics of Cortical Degeneration Over a Decade in Huntington's Disease. *Biological Psychiatry*, **2021**, 89, 807-816	7.9	10
61	In vivo neutralization of the protagonist role of macrophages during the chronic inflammatory stage of Huntington's disease. *Scientific Reports*, **2018**, 8, 11447	4.9	9
60	Quantification of huntingtin protein species in Huntington's disease patient leukocytes using optimised electrochemiluminescence immunoassays. *PLoS ONE*, **2017**, 12, e0189891	3.7	9
59	FAN1 controls mismatch repair complex assembly via MLH1 retention to stabilize CAG repeat expansion in Huntington's disease. *Cell Reports*, **2021**, 36, 109649	10.6	8
58	Inhibition of tumour necrosis factor alpha in the R6/2 mouse model of Huntington's disease by etanercept treatment. *Scientific Reports*, **2019**, 9, 7202	4.9	7
57	Disease Onset in Huntington's Disease: When Is the Conversion?. *Movement Disorders Clinical Practice*, **2021**, 8, 352-360	2.2	7
56	Composite UHDRS Correlates With Progression of Imaging Biomarkers in Huntington's Disease. *Movement Disorders*, **2021**, 36, 1259-1264	7	6
55	Reduction of confounding effects with voxel-wise Gaussian process regression in structural MRI **2014**,		5
54	Inflammatory markers in Huntington's disease plasma A robust nanoLC MRM-MS assay development. *EuPA Open Proteomics*, **2014**, 3, 68-75	0.1	5
53	Patients with Huntington's disease pioneered human stereotactic neurosurgery 70 years ago. *Brain*, **2017**, 140, 2516-2519	11.2	4

52	A MDS Evidence-Based Review on Treatments for Huntington's Disease. *Movement Disorders*, **2021**,	7	4
51	Premanifest and Early Huntington Disease **2014**,		4
50	Activity or Connectivity? Evaluating neurofeedback training in Huntington disease		4
49	Genetic testing in dementia - utility and clinical strategies. *Nature Reviews Neurology*, **2021**, 17, 23-36	15	4
48	. *Current Opinion in Neurology*, **2003**, 16, 451-458	7.1	3
47	Chapter 5 Mitochondrial Abnormalities in Neurodegenerative Disorders. *Blue Books of Practical Neurology*, **2002**, 26, 143-174		3
46	Fronto-striatal circuits for cognitive flexibility in far from onset Huntington's disease: evidence from the Young Adult Study. *Journal of Neurology, Neurosurgery and Psychiatry*, **2021**, 92, 143-149	5.5	3
45	F23 Validity, reliability, ability to detect change and meaningful within-patient change of the CUHDRS **2018**,		3
44	Longitudinal Structural MRI in Neurologically Healthy Adults. *Journal of Magnetic Resonance Imaging*, **2020**, 52, 1385-1399	5.6	2
43	Revealing the Timeline of Structural MRI Changes in Premanifest to Manifest Huntington Disease. *Neurology: Genetics*, **2021**, 7, e617	3.8	2
42	Longitudinal dynamics of mutant huntingtin and neurofilament light in Huntington disease: the prospective HD-CSF study		2
41	Magnetic Resonance Imaging in Huntington's Disease. *Methods in Molecular Biology*, **2018**, 1780, 303-328	1.4	2
40	A small molecule kicks repeat expansion into reverse. *Nature Genetics*, **2020**, 52, 136-137	36.3	1
39	QUANTIFYING MUTANT HUNTINGTIN IN HUNTINGTON'S DISEASE CSF. *Journal of Neurology, Neurosurgery and Psychiatry*, **2014**, 85, e4.132-e4	5.5	1
38	Relating quantitative 7T MRI across cortical depths to cytoarchitectonics, gene expression and connectomics: a framework for tracking neurodegenerative disease		1
37	Huntingtin lowering reduces somatic instability at CAG-expanded loci		1
36	Reply to 'Topographical layer imaging as a tool to track neurodegenerative disease spread in M1'. *Nature Reviews Neuroscience*, **2021**, 22, 69	13.5	1
35	Timing of selective basal ganglia white matter loss in Huntington disease		1

34	A Multi-Study Model-Based Evaluation of the Sequence of Imaging and Clinical Biomarker Changes in Huntington's Disease. *Frontiers in Big Data*, **2021**, 4, 662200	2.8	1
33	Huntington Disease Integrated Staging System (HD-ISS): A Novel Evidence-Based Classification System For Staging		1
32	Mislocalization of Nucleocytoplasmic Transport Proteins in Human Huntington's Disease PSC-Derived Striatal Neurons. *Frontiers in Cellular Neuroscience*, **2021**, 15, 742763	6.1	1
31	Huntington's disease64-82		0
30	Tracking Huntington's Disease Progression Using Motor, Functional, Cognitive, and Imaging Markers. *Movement Disorders*, **2021**, 36, 2282-2292	7	0
29	CAG Somatic Instability in a Huntington Disease Expansion Carrier Presenting with a Progressive Supranuclear Palsy-like Phenotype.. *Movement Disorders*, **2022**,	7	0
28	DNA REPAIR PATHWAYS MODULATE ONSET IN POLYGLUTAMINE DISEASES. *Journal of Neurology, Neurosurgery and Psychiatry*, **2016**, 87, e1.19-e1	5.5	
27	D20 Operationalising compensation over time in neurodegenerative disease. *Journal of Neurology, Neurosurgery and Psychiatry*, **2016**, 87, A41.2-A41	5.5	
26	B10 Inclusion formation in mutant HTT exon 1 expressing human neuronal cells. *Journal of Neurology, Neurosurgery and Psychiatry*, **2016**, 87, A12.2-A12	5.5	
25	D4 Prediction of huntington disease phenotype by cerebrospinal fluid biomarkers of inflammation and cell death. *Journal of Neurology, Neurosurgery and Psychiatry*, **2016**, 87, A35.1-A35	5.5	
24	D8 Tms-eeg markers of inhibitory deficits in huntington disease. *Journal of Neurology, Neurosurgery and Psychiatry*, **2016**, 87, A36.2-A36	5.5	
23	B24 Assessment of immune system activation status during the course of disease in huntington disease mouse model. *Journal of Neurology, Neurosurgery and Psychiatry*, **2016**, 87, A17.2-A17	5.5	
22	B15 Innate transcriptional dysregulation is associated with proinflammatory pathway activation in huntington disease myeloid cells. *Journal of Neurology, Neurosurgery and Psychiatry*, **2016**, 87, A14.1-A14	5.5	
21	D22 Compensation in preclinical huntington disease: evidence from the track-on HD study. *Journal of Neurology, Neurosurgery and Psychiatry*, **2016**, 87, A42.2-A42	5.5	
20	B25 Mitochondrial fission and fusion in skeletal muscle from HD patients and zQ175 mice. *Journal of Neurology, Neurosurgery and Psychiatry*, **2016**, 87, A17.3-A18	5.5	
19	J9 Probing huntington disease phenocopy syndromes with next-generation sequencing. *Journal of Neurology, Neurosurgery and Psychiatry*, **2016**, 87, A78.2-A78	5.5	
18	1609 Length of white matter connexions determine their rate of atrophy in premanifest huntington disease. *Journal of Neurology, Neurosurgery and Psychiatry*, **2017**, 88, A9.2-A9	5.5	
17	Opportunity cost determines free-operant action initiation latency and predicts apathy. *Psychological Medicine*,1-10	6.9	

16	Prion diseases of humans and animals **2010**, 243-250	
15	9 Aberrant striatal value representation in Huntington disease gene carriers 25 years before onset. *Journal of Neurology, Neurosurgery and Psychiatry*, **2020**, 91, e4.1-e4	5.5
14	D16 White matter microstructure and natural biological variation in huntington disease. *Journal of Neurology, Neurosurgery and Psychiatry*, **2016**, 87, A39.2-A39	5.5
13	B4 Detection of the aberrantly spliced exon 1 intron 1 htt mRNA in HD patient post mortem brain tissue and fibroblast lines. *Journal of Neurology, Neurosurgery and Psychiatry*, **2016**, 87, A10.2-A10	5.5
12	B27 Abnormal bioenergetics in inclusion-containing mutant HTT exon 1 primary human neurons. *Journal of Neurology, Neurosurgery and Psychiatry*, **2016**, 87, A18.2-A19	5.5
11	D18 Brain network breakdown and pathophysiological correlates in huntington disease. *Journal of Neurology, Neurosurgery and Psychiatry*, **2016**, 87, A40.2-A40	5.5
10	K4 The cost and value of a huntington disease multidisciplinary team meeting. *Journal of Neurology, Neurosurgery and Psychiatry*, **2016**, 87, A80.2-A80	5.5
9	D21 Longitudinal compensation in the cognitive network in huntington disease. *Journal of Neurology, Neurosurgery and Psychiatry*, **2016**, 87, A42.1-A42	5.5
8	B17 Blood transcriptome replicates dysregulation found in human huntington disease brain and shares an immune signature with alzheimer disease. *Journal of Neurology, Neurosurgery and Psychiatry*, **2016**, 87, A15.1-A15	5.5
7	B48 DNA repair pathways as a common genetic mechanism modulating the age at onset in polyglutamine diseases. *Journal of Neurology, Neurosurgery and Psychiatry*, **2016**, 87, A26.1-A26	5.5
6	D19 Longitudinal changes in functional connectivity of cortico-basal ganglia networks in manifest and premanifest huntington disease. *Journal of Neurology, Neurosurgery and Psychiatry*, **2016**, 87, A41.1-A41	5.5
5	G1 Executive task performance and anxiety are associated with self-awareness of neuropsychiatric symptoms in huntington disease. *Journal of Neurology, Neurosurgery and Psychiatry*, **2016**, 87, A54.2-A55	5.5
4	B49 Genetic modifiers of huntington disease progression. *Journal of Neurology, Neurosurgery and Psychiatry*, **2016**, 87, A26.2-A27	5.5
3	GENETIC MODIFIERS OF HUNTINGTON'S DISEASE PROGRESSION. *Journal of Neurology, Neurosurgery and Psychiatry*, **2016**, 87, e1.35-e1	5.5
2	Suppression of Somatic Expansion As a Novel Therapeutic Approach for Huntington Disease and Other Repeat Expansion Disorders **2022**, 1, 163-175	
1	241 Intrathecal antisense oligonucleotide delivery in HD: experience from RG6042 programme and best practice considerations. *Journal of Neurology, Neurosurgery and Psychiatry*, **2022**, 93, A83.1-A83	5.5