

# Jeffrey H Teckman

## List of Publications by Year in descending order

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116  
papers

4,734  
citations

94269

37  
h-index

102304

66  
g-index

123  
all docs

123  
docs citations

123  
times ranked

3503  
citing authors

#	ARTICLE	IF	CITATIONS
1	Degradation of a Mutant Secretory Protein, $\alpha_1$ -Antitrypsin Z, in the Endoplasmic Reticulum Requires Proteasome Activity. <i>Journal of Biological Chemistry</i> , 1996, 271, 22791-22795.	1.6	331
2	Retention of mutant $\alpha_1$ -antitrypsin Z in endoplasmic reticulum is associated with an autophagic response. <i>American Journal of Physiology - Renal Physiology</i> , 2000, 279, G961-G974.	1.6	244
3	Obese children with steatohepatitis can develop cirrhosis in childhood. <i>American Journal of Gastroenterology</i> , 2002, 97, 2460-2462.	0.2	224
4	$\alpha_1$ -Antitrypsin deficiency. <i>Nature Reviews Disease Primers</i> , 2016, 2, 16051.	18.1	215
5	Mitochondrial autophagy and injury in the liver in $\alpha_1$ -antitrypsin deficiency. <i>American Journal of Physiology - Renal Physiology</i> , 2004, 286, G851-G862.	1.6	183
6	The Diagnosis and Management of Alpha-1 Antitrypsin Deficiency in the Adult. <i>Chronic Obstructive Pulmonary Diseases (Miami, Fla )</i> , 2016, 3, 668-682.	0.5	148
7	A novel monoclonal antibody to characterize pathogenic polymers in liver disease associated with $\alpha_1$ -antitrypsin deficiency. <i>Hepatology</i> , 2010, 52, 1078-1088.	3.6	138
8	Defining the mechanism of polymerization in the serpinopathies. <i>Proceedings of the National Academy of Sciences of the United States of America</i> , 2010, 107, 17146-17151.	3.3	135
9	Gene transfer of master autophagy regulator TFEB results in clearance of toxic protein and correction of hepatic disease in $\alpha_1$ -antitrypsin deficiency. <i>EMBO Molecular Medicine</i> , 2013, 5, 397-412.	3.3	134
10	Molecular pathogenesis of liver disease in $\alpha_1$ -antitrypsin deficiency. <i>Hepatology</i> , 1996, 24, 1504-1516.	3.6	133
11	The Proteasome Participates in Degradation of Mutant $\alpha_1$ -Antitrypsin Z in the Endoplasmic Reticulum of Hepatoma-derived Hepatocytes. <i>Journal of Biological Chemistry</i> , 2001, 276, 44865-44872.	1.6	124
12	Analyses of hepatocellular proliferation in a mouse model of $\alpha_1$ -antitrypsin deficiency. <i>Hepatology</i> , 2004, 39, 1048-1055.	3.6	122
13	Rapamycin reduces intrahepatic alpha-1-antitrypsin mutant Z protein polymers and liver injury in a mouse model. <i>Experimental Biology and Medicine</i> , 2010, 235, 700-709.	1.1	110
14	Sustained miRNA-mediated Knockdown of Mutant AAT With Simultaneous Augmentation of Wild-type AAT Has Minimal Effect on Global Liver miRNA Profiles. <i>Molecular Therapy</i> , 2012, 20, 590-600.	3.7	105
15	Lack of Effect of Oral 4-Phenylbutyrate on Serum Alpha-1-Antitrypsin in Patients with $\alpha_1$ -Antitrypsin Deficiency: A Preliminary Study. <i>Journal of Pediatric Gastroenterology and Nutrition</i> , 2004, 39, 34-37.	0.9	97
16	Alpha-1-antitrypsin mutant Z protein content in individual hepatocytes correlates with cell death in a mouse model. <i>Hepatology</i> , 2007, 46, 1228-1235.	3.6	91
17	Fasting in $\alpha_1$ -antitrypsin deficient liver: constitutive activation of autophagy. <i>American Journal of Physiology - Renal Physiology</i> , 2002, 283, G1156-G1165.	1.6	80
18	The Endoplasmic Reticulum Degradation Pathway for Mutant Secretory Proteins $\alpha_1$ -Antitrypsin Z and S Is Distinct from That for an Unassembled Membrane Protein. <i>Journal of Biological Chemistry</i> , 1996, 271, 13215-13220.	1.6	78

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19	Diagnosis and Management of Patients With $\alpha$ -1-Antitrypsin (A1AT) Deficiency. <i>Clinical Gastroenterology and Hepatology</i> , 2012, 10, 575-580.	2.4	76
20	Liver Disease in Alpha-1 Antitrypsin Deficiency: Current Understanding and Future Therapy. <i>COPD: Journal of Chronic Obstructive Pulmonary Disease</i> , 2013, 10, 35-43.	0.7	71
21	Antisense oligonucleotide treatment ameliorates alpha-1 antitrypsin-related liver disease in mice. <i>Journal of Clinical Investigation</i> , 2014, 124, 251-261.	3.9	70
22	A Naturally Occurring Nonpolymerogenic Mutant of $\alpha$ -1-Antitrypsin Characterized by Prolonged Retention in the Endoplasmic Reticulum. <i>Journal of Biological Chemistry</i> , 2001, 276, 33893-33898.	1.6	60
23	Quantitative isolation of $\alpha$ 1AT mutant Z protein polymers from human and mouse livers and the effect of heat. <i>Hepatology</i> , 2005, 41, 160-167.	3.6	58
24	Alpha-1-antitrypsin deficiency: Genetic variations, clinical manifestations and therapeutic interventions. <i>Mutation Research - Reviews in Mutation Research</i> , 2017, 773, 14-25.	2.4	58
25	In vivo post-transcriptional gene silencing of $\alpha$ -1 antitrypsin by adeno-associated virus vectors expressing siRNA. <i>Laboratory Investigation</i> , 2007, 87, 893-902.	1.7	57
26	Role of ubiquitin in proteasomal degradation of mutant $\alpha$ -1-antitrypsin Z in the endoplasmic reticulum. <i>American Journal of Physiology - Renal Physiology</i> , 2000, 278, G39-G48.	1.6	55
27	Indomethacin increases liver damage in a murine model of liver injury from alpha-1-antitrypsin deficiency. <i>Hepatology</i> , 2006, 44, 976-982.	3.6	55
28	Determination of hepatitis B phenotype using biochemical and serological markers. <i>Journal of Viral Hepatitis</i> , 2017, 24, 320-329.	1.0	55
29	Alpha-1-Antitrypsin Deficiency Liver Disease. <i>Clinics in Liver Disease</i> , 2018, 22, 643-655.	1.0	54
30	Oxidative stress contributes to liver damage in a murine model of alpha-1-antitrypsin deficiency. <i>Experimental Biology and Medicine</i> , 2012, 237, 1163-1172.	1.1	51
31	Amelioration of Alpha-1 Antitrypsin Deficiency Diseases with Genome Editing in Transgenic Mice. <i>Human Gene Therapy</i> , 2018, 29, 861-873.	1.4	49
32	$\alpha$ -1-Antitrypsin Deficiency in Childhood. <i>Seminars in Liver Disease</i> , 2007, 27, 274-281.	1.8	47
33	Combination of Entecavir/Peginterferon Alfa-2a in Children With Hepatitis B e Antigen-Positive Immune Tolerant Chronic Hepatitis B Virus Infection. <i>Hepatology</i> , 2019, 69, 2326-2337.	3.6	45
34	Alpha-1 antitrypsin and liver disease: mechanisms of injury and novel interventions. <i>Expert Review of Gastroenterology and Hepatology</i> , 2015, 9, 261-268.	1.4	44
35	Corneal opacities associated with NTBC treatment. <i>American Journal of Ophthalmology</i> , 2002, 134, 266-268.	1.7	41
36	Baseline Analysis of a Young $\alpha$ -1-Antitrypsin Deficiency Liver Disease Cohort Reveals Frequent Portal Hypertension. <i>Journal of Pediatric Gastroenterology and Nutrition</i> , 2015, 61, 94-101.	0.9	41

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37	Activating Transcription Factor 6 Limits Intracellular Accumulation of Mutant $\alpha$ 1-Antitrypsin Z and Mitochondrial Damage in Hepatoma Cells. <i>Journal of Biological Chemistry</i> , 2011, 286, 41563-41577.	1.6	40
38	Neurodevelopmental Outcome of Young Children with Biliary Atresia and Native Liver: Results from the ChiLDRen Study. <i>Journal of Pediatrics</i> , 2018, 196, 139-147.e3.	0.9	40
39	REVIEW: $\alpha$ 1-Antitrypsin deficiency associated liver disease. <i>Journal of Gastroenterology and Hepatology (Australia)</i> , 1997, 12, 404-416.	1.4	39
40	Characteristics of hepatocellular carcinoma in a murine model of $\alpha$ 1-antitrypsin deficiency. <i>Hepatology Research</i> , 2010, 40, 641-653.	1.8	38
41	Alpha-1-antitrypsin deficiency: Diagnosis, pathophysiology, and management. <i>Current Gastroenterology Reports</i> , 2006, 8, 14-20.	1.1	37
42	Development of an RNAi therapeutic for alpha-1-antitrypsin liver disease. <i>JCI Insight</i> , 2020, 5, .	2.3	37
43	Sustained Knockdown of a Disease-Causing Gene in Patient-Specific Induced Pluripotent Stem Cells Using Lentiviral Vector-Based Gene Therapy. <i>Stem Cells Translational Medicine</i> , 2013, 2, 641-654.	1.6	36
44	The Serpinopathies. <i>Methods in Enzymology</i> , 2011, 501, 421-466.	0.4	35
45	Advances in Alpha-1-Antitrypsin Deficiency Liver Disease. <i>Current Gastroenterology Reports</i> , 2014, 16, 367.	1.1	32
46	Bone Density in Children With Chronic Liver Disease Correlates With Growth and Cholestasis. <i>Hepatology</i> , 2019, 69, 245-257.	3.6	31
47	$\alpha$ 1-Antitrypsin Deficiency: From Genotype to Childhood Disease. <i>Journal of Pediatric Gastroenterology and Nutrition</i> , 1998, 27, 65-74.	0.9	30
48	Molecular pathogenesis of liver disease in alpha1-antitrypsin deficiency. <i>Hepatology</i> , 1996, 24, 1504-1516.	3.6	29
49	Children with Chronic Hepatitis B in the United States and Canada. <i>Journal of Pediatrics</i> , 2015, 167, 1287-1294.e2.	0.9	28
50	Autophagy induced by exogenous bile acids is therapeutic in a model of $\alpha$ 1-AT deficiency liver disease. <i>American Journal of Physiology - Renal Physiology</i> , 2016, 311, G156-G165.	1.6	27
51	NorUDCA promotes degradation of $\alpha$ 1-antitrypsin mutant Z protein by inducing autophagy through AMPK/ULK1 pathway. <i>PLoS ONE</i> , 2018, 13, e0200897.	1.1	27
52	Conceptual advances in the pathogenesis and treatment of childhood metabolic liver disease. <i>Gastroenterology</i> , 1995, 108, 1263-1279.	0.6	25
53	Quality of Life and Its Determinants in a Multicenter Cohort of Children with Alagille Syndrome. <i>Journal of Pediatrics</i> , 2015, 167, 390-396.e3.	0.9	25
54	Bone marrow stem cell therapy partially ameliorates pathological consequences in livers of mice expressing mutant human $\alpha$ 1-antitrypsin. <i>Hepatology</i> , 2017, 65, 1319-1335.	3.6	25

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55	Pathophysiology of Alpha-1 Antitrypsin Deficiency Liver Disease. <i>Methods in Molecular Biology</i> , 2017, 1639, 1-8.	0.4	25
56	Hepatic Progenitor Cell Proliferation and Liver Injury in $\alpha$ -1 Antitrypsin Deficiency. <i>Journal of Pediatric Gastroenterology and Nutrition</i> , 2010, 51, 626-630.	0.9	24
57	Total Serum Bilirubin Predicts Fat-Soluble Vitamin Deficiency Better Than Serum Bile Acids in Infants With Biliary Atresia. <i>Journal of Pediatric Gastroenterology and Nutrition</i> , 2014, 59, 702-707.	0.9	24
58	Activation of the c-Jun N-terminal kinase pathway aggravates proteotoxicity of hepatic mutant Z $\alpha$ -1 antitrypsin. <i>Hepatology</i> , 2017, 65, 1865-1874.	3.6	24
59	Gastrointestinal pathophysiology and nutrition in cystic fibrosis. <i>Expert Review of Gastroenterology and Hepatology</i> , 2018, 12, 853-862.	1.4	24
60	Controversies in the Mechanism of Total Parenteral Nutrition Induced Pathology. <i>Children</i> , 2015, 2, 358-370.	0.6	23
61	Preserved Gut Microbial Diversity Accompanies Upregulation of TGR5 and Hepatobiliary Transporters in Bile Acid-Treated Animals Receiving Parenteral Nutrition. <i>Journal of Parenteral and Enteral Nutrition</i> , 2017, 41, 198-207.	1.3	22
62	Oleanolic Acid Improves Gut Atrophy Induced by Parenteral Nutrition. <i>Journal of Parenteral and Enteral Nutrition</i> , 2016, 40, 67-72.	1.3	21
63	Appropriateness of Newborn Screening for $\alpha$ -1 Antitrypsin Deficiency. <i>Journal of Pediatric Gastroenterology and Nutrition</i> , 2014, 58, 199-203.	0.9	20
64	Alpha-1 antitrypsin deficiency liver disease. <i>Translational Gastroenterology and Hepatology</i> , 2021, 6, 23-23.	1.5	20
65	Detection of alpha-1 antitrypsin deficiency: the past, present and future. <i>Orphanet Journal of Rare Diseases</i> , 2020, 15, 96.	1.2	18
66	Review of Gastrointestinal Motility in Cystic Fibrosis. <i>Journal of Cystic Fibrosis</i> , 2021, 20, 578-585.	0.3	18
67	CHOP and c-JUN up-regulate the mutant Z $\alpha$ -1-antitrypsin, exacerbating its aggregation and liver proteotoxicity. <i>Journal of Biological Chemistry</i> , 2020, 295, 13213-13223.	1.6	16
68	Up-regulation of miR-34b/c by JNK and FOXO3 protects from liver fibrosis. <i>Proceedings of the National Academy of Sciences of the United States of America</i> , 2021, 118, .	3.3	16
69	Validating hyperbilirubinemia and gut mucosal atrophy with a novel ultramobile ambulatory total parenteral nutrition piglet model. <i>Nutrition Research</i> , 2015, 35, 169-174.	1.3	15
70	Pediatric Intestinal Failure Review. <i>Children</i> , 2018, 5, 100.	0.6	14
71	Phenotypes of Chronic Hepatitis B in Children From a Large North American Cohort. <i>Journal of Pediatric Gastroenterology and Nutrition</i> , 2019, 69, 588-594.	0.9	13
72	Chronic Hepatitis Is Common and Often Untreated Among Children with Hepatitis B Infection in the United States and Canada. <i>Journal of Pediatrics</i> , 2021, 237, 24-33.e12.	0.9	10

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73	Emerging Concepts and Human Trials in Alpha-1-Antitrypsin Deficiency Liver Disease. <i>Seminars in Liver Disease</i> , 2017, 37, 152-158.	1.8	9
74	Longitudinal Outcomes in Young Patients with Alpha-1-Antitrypsin Deficiency with Native Liver Reveal that Neonatal Cholestasis is a Poor Predictor of Future Portal Hypertension. <i>Journal of Pediatrics</i> , 2020, 227, 81-86.e4.	0.9	9
75	Diagnostic accuracy of non-contrast magnetic resonance enterography in detecting active bowel inflammation in pediatric patients with diagnosed or suspected inflammatory bowel disease to determine necessity of gadolinium-based contrast agents. <i>Pediatric Radiology</i> , 2019, 49, 759-769.	1.1	8
76	Liver disease with unknown etiology “ have you ruled out alpha-1 antitrypsin deficiency?. <i>Therapeutic Advances in Chronic Disease</i> , 2021, 12_suppl, 204062232199568.	1.1	8
77	279 Pre-Clinical Evaluation of ALN-AAT to Ameliorate Liver Disease Associated With Alpha-1-Antitrypsin Deficiency. <i>Gastroenterology</i> , 2015, 148, S-975.	0.6	7
78	AFM Imaging Reveals Topographic Diversity of Wild Type and Z Variant Polymers of Human $\alpha$ 1-Proteinase Inhibitor. <i>PLoS ONE</i> , 2016, 11, e0151902.	1.1	7
79	Health-related Quality of Life in Pediatric Patients With Chronic Hepatitis B Living in the United States and Canada. <i>Journal of Pediatric Gastroenterology and Nutrition</i> , 2017, 64, 760-769.	0.9	7
80	Clinical Trial Design for Alpha-1 Antitrypsin Deficiency: A Model for Rare Diseases. <i>Chronic Obstructive Pulmonary Diseases (Miami, Fla )</i> , 2014, 2, 177-190.	0.5	7
81	Changes in serum hepatitis B surface and e antigen, interferon-inducible protein 10, and aminotransferase levels during combination therapy of immune-tolerant chronic hepatitis B. <i>Hepatology</i> , 2022, 76, 775-787.	3.6	7
82	Alpha-1 Antitrypsin Deficiency Liver Disease. <i>Clinics in Liver Disease</i> , 2022, 26, 391-402.	1.0	7
83	PiZ Mouse Liver Accumulates Polyubiquitin Conjugates That Associate with Catalytically Active 26S Proteasomes. <i>PLoS ONE</i> , 2014, 9, e106371.	1.1	6
84	Hepatic Histology in Treatment-naïve Children With Chronic Hepatitis B Infection Living in the United States and Canada. <i>Journal of Pediatric Gastroenterology and Nutrition</i> , 2020, 71, 99-105.	0.9	6
85	Hepatitis B e antigen loss in adults and children with chronic hepatitis B living in North America: A prospective cohort study. <i>Journal of Viral Hepatitis</i> , 2021, 28, 1526-1538.	1.0	6
86	Clinical significance of quantitative e antigen in a cohort of hepatitis B virus-infected children and adults in North America. <i>Journal of Viral Hepatitis</i> , 2021, 28, 1042-1056.	1.0	5
87	Author's Response. <i>Journal of Pediatric Gastroenterology and Nutrition</i> , 2015, 60, e38.	0.9	4
88	Leveraging Population Genomics for Individualized Correction of the Hallmarks of Alpha-1 Antitrypsin Deficiency. <i>Chronic Obstructive Pulmonary Diseases (Miami, Fla )</i> , 2020, 7, 224-246.	0.5	4
89	Semiquantitation of Monomer and Polymer Alpha-1 Antitrypsin by Centrifugal Separation and Assay by Western Blot of Soluble and Insoluble Components. <i>Methods in Molecular Biology</i> , 2017, 1639, 227-234.	0.4	3
90	Alpha-1- Antitrypsin Deficiency Liver Disease: Science and Therapeutic Potential 50 Years Later. <i>Journal of Gastroenterology, Pancreatology &amp; Liver Disorders</i> , 2014, 1, .	0.2	3

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91	Liver Disease Due to Alpha-1 Antitrypsin Deficiency: Are We Surprised That It Is More Complex Than We Thought?. <i>Hepatology</i> , 2019, 70, 5-7.	3.6	2
92	Clinical approach to paediatric liver disease. , 2019, , 105-113.		2
93	Alpha-1 Antitrypsin Deficiency. <i>Clinical Liver Disease</i> , 2022, 19, 89-92.	1.0	2
94	593 Antisense Oligonucleotide Reduction of Mutant Alpha-1 Antitrypsin Protein for the Treatment of Alpha-1 Antitrypsin Liver Disease. <i>Gastroenterology</i> , 2012, 142, S-919.	0.6	1
95	Review of Treatment Outcomes in Pediatric Patients with Concomitant Eosinophilic Esophagitis and Celiac Disease. <i>Journal of Allergy and Clinical Immunology</i> , 2013, 131, AB181.	1.5	1
96	Author's Response. <i>Journal of Pediatric Gastroenterology and Nutrition</i> , 2015, 61, e26.	0.9	1
97	Alpha-1-Antitrypsin Deficiency Liver Disease. , 2017, , 117-131.		1
98	Novel NMP split liver model recapitulates human IRI and demonstrates ferroptosis modulators as a new therapeutic strategy. <i>Pediatric Transplantation</i> , 2021, , e14164.	0.5	1
99	Hepatocellular proliferation in a mouse model of alpha-1-antitrypsin deficiency. <i>Gastroenterology</i> , 2003, 124, A713.	0.6	0
100	Isolation of alpha-1-antitrypsin mutant Z protein polymers from liver and their role in cellular injury. <i>Gastroenterology</i> , 2003, 124, A689.	0.6	0
101	Selected Summary. <i>Journal of Pediatric Gastroenterology and Nutrition</i> , 2009, 48, 115-116.	0.9	0
102	Pro-Inflammatory Consequences Of Human Z Protein After Cigarette Smoke Exposure. , 2011, , .		0
103	702 Baseline Analysis of the Largest Reported Cohort of Children and Young Adults With Alpha-1-Antitrypsin Deficiency Liver Disease Reveals a Significant Burden of Portal Hypertension Without Jaundice. <i>Gastroenterology</i> , 2012, 142, S-924.	0.6	0
104	850c RNAi Therapeutics Ameliorate Liver Disease Associated with Alpha-1-Antitrypsin Deficiency. <i>Gastroenterology</i> , 2014, 146, S-145.	0.6	0
105	Tu1127 Health-Related Quality of Life in a Cohort of Children With Chronic Hepatitis B. <i>Gastroenterology</i> , 2014, 146, S-761.	0.6	0
106	Mo1679 Development and Validation of a Novel Ultra-Mobile Ambulatory Total Parenteral Nutrition Model. <i>Gastroenterology</i> , 2014, 146, S-634.	0.6	0
107	Su1004 Drug Resistance, Basal Core Promoter and Pre-Core Stop Mutations in a Large Cohort of North American Children With Hepatitis B: Results for the Pediatric Cohort Study of the NIDDK-Sponsored Hepatitis B Research Network (HBRN). <i>Gastroenterology</i> , 2014, 146, S-960.	0.6	0
108	Su1798 Glucagon-Like Peptides Ameliorate Total Prenteral Nutrition Associated Gut Atrophy. <i>Gastroenterology</i> , 2015, 148, S-1054.	0.6	0

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109	808 HBeAg(+) Immune Tolerant Phenotype Is Rare Among Children In The Largest US-Canadian Cohort With Chronic HBV Infection. <i>Gastroenterology</i> , 2016, 150, S1048.	0.6	0
110	940 Death Is Rare and Progression to Transplant Is Slow, Despite a Burden of Portal Hypertension, in Longitudinal Outcomes of Alpha-1-Antitrypsin Deficiency From the Children Cohort. <i>Gastroenterology</i> , 2016, 150, S1052-S1053.	0.6	0
111	Quantitative Surface Antigen and E Antigen in Children with Chronic Hepatitis B: Results from the Pediatric Cohort Study of the Niddk-Sponsored Hepatitis B Research Network (HBRN). <i>Gastroenterology</i> , 2017, 152, S1071-S1072.	0.6	0
112	Alpha-1 Antitrypsin Deficiency: Cytotoxic Z Polymer Formation in the Lungs Promotes Increased Apoptosis. , 2020, , .		0
113	Alpha-1-Antitrypsin ( $\hat{\pm}$ 1AT) Deficiency, <i>Pediatric.</i> , 2004, , 48-51.		0
114	Alpha-1-Antitrypsin Deficiency and Mechanisms of Liver Disease. <i>Anti-Inflammatory and Anti-Allergy Agents in Medicinal Chemistry</i> , 2010, 9, 289-298.	1.1	0
115	Alpha-1-Antitrypsin Deficiency Liver Disease and New Treatment Opportunities. <i>Anti-Inflammatory and Anti-Allergy Agents in Medicinal Chemistry</i> , 1970, 10, 382-391.	1.1	0
116	Innovations in Pediatrics: 2019. <i>Missouri Medicine</i> , 2019, 116, 105.	0.3	0