

# Richard J Thompson

## List of Publications by Year in descending order

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Version: 2024-02-01

88  
papers

6,350  
citations

109137

35  
h-index

66788

78  
g-index

93  
all docs

93  
docs citations

93  
times ranked

3915  
citing authors

| #  | ARTICLE  | IF  | CITATIONS |
|----|--|-----|-----------|
| 1  | Liver Disease and Risk of Hepatocellular Carcinoma in Children With Mutations in TALDO1. <i>Hepatology Communications</i> , 2022, 6, 473-479.  | 2.0 | 8         |
| 2  | Human iPSC-derived hepatocyte system models cholestasis with tight junction protein 2 deficiency. <i>JHEP Reports</i> , 2022, 4, 100446.   | 2.6 | 5         |
| 3  | Maralixibat for the treatment of PFIC: Long-term, IBAT inhibition in an open-label, Phase 2 study. <i>Hepatology Communications</i> , 2022, 6, 2379-2390.  | 2.0 | 26        |
| 4  | Impact of long-term administration of maralixibat on children with cholestasis secondary to Alagille syndrome. <i>Hepatology Communications</i> , 2022, 6, 1922-1933.                                      | 2.0 | 12        |
| 5  | Complications of congenital portosystemic shunts: liver tumors are affected by shunt severity, but pulmonary and neurocognitive associations are not. <i>Hepatology International</i> , 2022, 16, 918-925. | 1.9 | 5         |
| 6  | Odevixibat treatment in progressive familial intrahepatic cholestasis: a randomised, placebo-controlled, phase 3 trial. <i>The Lancet Gastroenterology and Hepatology</i> , 2022, 7, 830-842.              | 3.7 | 54        |
| 7  | Challenges in understanding the consequences of variants in ABCB4 gene. <i>Journal of Hepatology</i> , 2021, 74, 242-243.  | 1.8 | 1         |
| 8  | Use of a Comprehensive 66-Gene Cholestasis Sequencing Panel in 2171 Cholestatic Infants, Children, and Young Adults. <i>Journal of Pediatric Gastroenterology and Nutrition</i> , 2021, 72, 654-660.       | 0.9 | 20        |
| 9  | The Influence of Donor and Recipient Complement C3 Polymorphisms on Liver Transplant Outcome. <i>International Journal of Hepatology</i> , 2021, 2021, 1-14.   | 0.4 | 1         |
| 10 | Progressive familial intrahepatic cholestasis " farnesoid X receptor deficiency due to <i>NR1H4</i> mutation: A case report. <i>World Journal of Clinical Cases</i> , 2021, 9, 3631-3636.                  | 0.3 | 7         |
| 11 | Mutation Analysis and Disease Features at Presentation in a Multi-Center Cohort of Children With Monogenic Cholestasis. <i>Journal of Pediatric Gastroenterology and Nutrition</i> , 2021, 73, 169-177.    | 0.9 | 8         |
| 12 | Impact of Genotype, Serum Bile Acids, and Surgical Biliary Diversion on Native Liver Survival in FIC1 Deficiency. <i>Hepatology</i> , 2021, 74, 892-906.   | 3.6 | 25        |
| 13 | Effects of odevixibat on pruritus and bile acids in children with cholestatic liver disease: Phase 2 study. <i>Clinics and Research in Hepatology and Gastroenterology</i> , 2021, 45, 101751.             | 0.7 | 40        |
| 14 | Study of Acute Liver Failure in Children Using Next Generation Sequencing Technology. <i>Journal of Pediatrics</i> , 2021, 236, 124-130.   | 0.9 | 7         |
| 15 | Cholestasis Due to USP53 Deficiency. <i>Journal of Pediatric Gastroenterology and Nutrition</i> , 2021, 72, 667-673.   | 0.9 | 18        |
| 16 | Portal venous deprivation in patients with portosystemic shunts and its effect on liver tumors. <i>Journal of Pediatric Surgery</i> , 2020, 55, 651-654.   | 0.8 | 5         |
| 17 | Mutations in Myosin 5B in Children With Early-onset Cholestasis. <i>Journal of Pediatric Gastroenterology and Nutrition</i> , 2020, 71, 184-188.   | 0.9 | 16        |
| 18 | Enriched conditioning expands the regenerative ability of sensory neurons after spinal cord injury via neuronal intrinsic redox signaling. <i>Nature Communications</i> , 2020, 11, 6425.                  | 5.8 | 37        |

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|----|---|-----|-----------|
| 19 | 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         |
| 20 | Systematic Review and Meta-analysis. <i>Journal of Pediatric Gastroenterology and Nutrition</i> , 2020, 71, 176-183.  | 0.9 | 21        |
| 21 | Genotype correlates with the natural history of severe bile salt export pump deficiency. <i>Journal of Hepatology</i> , 2020, 73, 84-93.  | 1.8 | 61        |
| 22 | Review article: liver disease in adults with variants in the cholestasis-related genes <i>ABCB11</i> , <i>ABCB4</i> and <i>ATP8B1</i> . <i>Alimentary Pharmacology and Therapeutics</i> , 2020, 52, 1628-1639.                                    | 1.9 | 25        |
| 23 | Biliary transporter gene mutations in severe intrahepatic cholestasis of pregnancy: Diagnostic and management implications. <i>Journal of Gastroenterology and Hepatology (Australia)</i> , 2019, 34, 425-435.                                    | 1.4 | 8         |
| 24 | Identification of Polycystic Kidney Disease 1 Like 1 Gene Variants in Children With Biliary Atresia Splenic Malformation Syndrome. <i>Hepatology</i> , 2019, 70, 899-910.   | 3.6 | 58        |
| 25 | Histopathology of livers in patients with congenital portosystemic shunts (Abernethy malformation): a case series of 22 patients. <i>Virchows Archiv Fur Pathologische Anatomie Und Physiologie Und Fur Klinische Medizin</i> , 2019, 474, 47-57. | 1.4 | 24        |
| 26 | Outcomes of surgical management of familial intrahepatic cholestasis 1 and bile salt export protein deficiencies. <i>Hepatology Communications</i> , 2018, 2, 515-528.  | 2.0 | 54        |
| 27 | Massive ascites and the heterozygous alpha 1 antitrypsin ( $\alpha_1$ -AT) living related donor liver in the homozygous child. <i>Pediatric Transplantation</i> , 2018, 22, e13075.   | 0.5 | 5         |
| 28 | Reduced Hepatocellular Expression of Canalicular Transport Proteins in Infants with Neonatal Cholestasis and Congenital Hypopituitarism. <i>Journal of Pediatrics</i> , 2018, 200, 181-187.   | 0.9 | 10        |
| 29 | Progressive Familial Intrahepatic Cholestasis. <i>Clinics in Liver Disease</i> , 2018, 22, 657-669.   | 1.0 | 124       |
| 30 | Donor transmitted mutation of the <i>ABCB11</i> gene and ensuing intrahepatic cholestasis of pregnancy in a liver transplant recipient. <i>Liver Transplantation</i> , 2017, 23, 1229-1232.   | 1.3 | 2         |
| 31 | An expanded role for heterozygous mutations of <i>ABCB4</i> , <i>ABCB11</i> , <i>ATP8B1</i> , <i>ABCC2</i> and <i>TJP2</i> in intrahepatic cholestasis of pregnancy. <i>Scientific Reports</i> , 2017, 7, 11823.                                  | 1.6 | 98        |
| 32 | Sequencing of transporter genes in cholestasis: We are still learning. <i>Journal of Hepatology</i> , 2017, 67, 1132-1133.  | 1.8 | 3         |
| 33 | Sequencing of <i>FIC1</i> , <i>BSEP</i> and <i>MDR3</i> in a large cohort of patients with cholestasis revealed a high number of different genetic variants. <i>Journal of Hepatology</i> , 2017, 67, 1253-1264.                                  | 1.8 | 97        |
| 34 | Reply to: "Doublecortin domain containing protein 2 (DCDC2) genetic variants in primary sclerosing cholangitis". <i>Journal of Hepatology</i> , 2017, 67, 652-653.  | 1.8 | 0         |
| 35 | Multiple $\beta$ -catenin mutations in hepatocellular lesions arising in Abernethy malformation. <i>Human Pathology</i> , 2016, 53, 153-158.  | 1.1 | 32        |
| 36 | Mutations in <i>DCDC2</i> (doublecortin domain containing protein 2) in neonatal sclerosing cholangitis. <i>Journal of Hepatology</i> , 2016, 65, 1179-1187.  | 1.8 | 65        |

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|----|---|-----|-----------|
| 37 | Familial Intrahepatic Cholestasis. , 2016, , 663-670.   |     | 0         |
| 38 | Hepatocellular carcinoma associated with tightâ€junction protein 2 deficiency. Hepatology, 2015, 62, 1914-1916.  | 3.6 | 63        |
| 39 | Pigmented well-differentiated hepatocellular neoplasm with $\beta$ -catenin mutation. Hepatobiliary and Pancreatic Diseases International, 2015, 14, 660-664.   | 0.6 | 5         |
| 40 | Antiâ€CD20 Monoclonal Antibody Therapy in Functional Bile Salt Export Pump Deficiency After Liver Transplantation. Journal of Pediatric Gastroenterology and Nutrition, 2015, 60, e50-3.                           | 0.9 | 6         |
| 41 | Genetics of liver disease: From pathophysiology to clinical practice. Journal of Hepatology, 2015, 62, S6-S14.  | 1.8 | 73        |
| 42 | Treating genetic disease: Expanding the options. Hepatology, 2015, 62, 349-351.   | 3.6 | 4         |
| 43 | Mutations in <i>TJP2</i> , encoding zona occludens 2, and liver disease. Tissue Barriers, 2015, 3, e1026537.  | 1.6 | 55        |
| 44 | Massive gene amplification drives paediatric hepatocellular carcinoma caused by bile salt export pump deficiency. Nature Communications, 2014, 5, 3850.   | 5.8 | 49        |
| 45 | Microvilli as markers of disordered apical-membrane trafficking and assembly: Bowel and liver. Hepatology, 2014, 60, 34-36.   | 3.6 | 6         |
| 46 | Mutations in TJP2 cause progressive cholestatic liver disease. Nature Genetics, 2014, 46, 326-328.  | 9.4 | 244       |
| 47 | Genetic and metabolic liver disease. , 2012, , 157-259.   |     | 8         |
| 48 | ATP8B1 Gene Expression Is Driven by a Housekeeping-Like Promoter Independent of Bile Acids and Farnesoid X Receptor. PLoS ONE, 2012, 7, e51650.   | 1.1 | 4         |
| 49 | Recurrent low gamma-glutamyl Transpeptidase cholestasis following liver transplantation for bile salt export pump (BSEP) disease (posttransplant recurrent BSEP disease). Liver Transplantation, 2010, 16, 856-863. | 1.3 | 60        |
| 50 | Differences in presentation and progression between severe FIC1 and BSEP deficiencies. Journal of Hepatology, 2010, 53, 170-178.  | 1.8 | 182       |
| 51 | Contribution of variant alleles of ABCB11 to susceptibility to intrahepatic cholestasis of pregnancy. Gut, 2009, 58, 537-544.   | 6.1 | 179       |
| 52 | Missense mutations and single nucleotide polymorphisms in ABCB11 impair bile salt export pump processing and function or disrupt pre-messenger RNA splicing. Hepatology, 2009, 49, 553-567.                         | 3.6 | 147       |
| 53 | Effect of ischemic preconditioning on the genomic response to reperfusion injury in deceased donor liver transplantation. Liver Transplantation, 2009, 15, 1750-1765.   | 1.3 | 36        |
| 54 | Severe bile salt export pump (BSEP) deficiency: mutations, immunohistochemically assessed BSEP expression, and malignancy risk. , 2009, , 206-216.  |     | 0         |

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|----|---|-----|-----------|
| 55 | The role of ABCB11 (BSEP) variation in susceptibility to intrahepatic cholestasis of pregnancy. , 2009, , 46-50.  |     | 0         |
| 56 | Bile composition in Alagille Syndrome and PFIC patients having Partial External Biliary Diversion. BMC Gastroenterology, 2008, 8, 47.   | 0.8 | 25        |
| 57 | Healy's law and the pediatric liver allograft. Liver Transplantation, 2008, 14, 1557-1558.  | 1.3 | 1         |
| 58 | Severe Bile Salt Export Pump Deficiency: 82 Different ABCB11 Mutations in 109 Families. Gastroenterology, 2008, 134, 1203-1214.e8.  | 0.6 | 331       |
| 59 | Congenital bile duct anomalies (biliary atresia) and chromosome 22 aneuploidy. Journal of Pediatric Surgery, 2008, 43, 1736-1740.   | 0.8 | 38        |
| 60 | 879 CHOLANGIOPATHY AND HCC IN MDR3 DEFICIENCY. Journal of Hepatology, 2008, 48, S330.   | 1.8 | 0         |
| 61 | S ndromes colest ticos familiares. Annales Nestl  (Ed Espa ola), 2008, 66, 121-126.   | 0.1 | 0         |
| 62 | Familial Cholestatic Syndromes. Annales Nestle, 2008, 66, 121-126.  | 0.1 | 0         |
| 63 | Cholestasis: Current Issues and Plan for the Future. Journal of Pediatric Gastroenterology and Nutrition, 2008, 47, 220-224.  | 0.9 | 9         |
| 64 | Syndromes cholestatiques familiaux. Annales Nestle [Ed Francaise], 2008, 66, 121-126.   | 0.0 | 0         |
| 65 | Lack of hepatocellular CD10 along bile canaliculi is physiologic in early childhood and persistent in Alagille syndrome. Laboratory Investigation, 2007, 87, 1138-1148.             | 1.7 | 31        |
| 66 | Mutations in Bile Salt Export Pump (ABCB11) in Two Children with Progressive Familial Intrahepatic Cholestasis and Cholangiocarcinoma. Journal of Pediatrics, 2007, 150, 556-559.   | 0.9 | 129       |
| 67 | VPS33B mutation with ichthyosis, cholestasis, and renal dysfunction but without arthrogyrosis: Incomplete ARC syndrome phenotype. Journal of Pediatrics, 2006, 148, 269-271.        | 0.9 | 50        |
| 68 | Exocrine Pancreatic Function in Children with Progressive Familial Intrahepatic Cholestasis Type 2. Journal of Pediatric Gastroenterology and Nutrition, 2006, 42, 416-418.         | 0.9 | 20        |
| 69 | Hepatocellular carcinoma in ten children under five years of age with bile salt export pump deficiency. Hepatology, 2006, 44, 478-486.  | 3.6 | 345       |
| 70 | Heterogeneity of the egammadeltabeta-thalassaemias: characterization of three novel English deletions. British Journal of Haematology, 2005, 128, 722-729.                          | 1.2 | 35        |
| 71 | Mutations in VPS33B, encoding a regulator of SNARE-dependent membrane fusion, cause arthrogyrosis renal dysfunction cholestasis (ARC) syndrome. Nature Genetics, 2004, 36, 400-404. | 9.4 | 313       |
| 72 | Characterization of mutations inATP8B1associated with hereditary cholestasis. Hepatology, 2004, 40, 27-38.  | 3.6 | 263       |

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|----|---|-----|-----------|
| 73 | Progressive familial intrahepatic cholestasis, type 1, is associated with decreased farnesoid X receptor activity. <i>Gastroenterology</i> , 2004, 126, 756-764.  | 0.6 | 185       |
| 74 | Cholestatic and Metabolic Liver Diseases: Working Group Report of the Second World Congress of Pediatric Gastroenterology, Hepatology, and Nutrition. <i>Journal of Pediatric Gastroenterology and Nutrition</i> , 2004, 39, S611-S615.   | 0.9 | 11        |
| 75 | Novel Mutations Responsible for Autosomal Recessive Multisystem Pseudohypoaldosteronism and Sequence Variants in Epithelial Sodium Channel $\beta_1$ , $\beta_2$ , and $\beta_3$ -Subunit Genes. <i>Journal of Clinical Endocrinology and Metabolism</i> , 2002, 87, 3344-3350. | 1.8 | 48        |
| 76 | The human bile salt export pump: Characterization of substrate specificity and identification of inhibitors. <i>Gastroenterology</i> , 2002, 123, 1649-1658.  | 0.6 | 308       |
| 77 | BSEP: Function and Role in Progressive Familial Intrahepatic Cholestasis. <i>Seminars in Liver Disease</i> , 2001, 21, 545-550.   | 1.8 | 117       |
| 78 | Pediatric liver disease. <i>Current Opinion in Gastroenterology</i> , 2000, 16, 271-274.  | 1.0 | 1         |
| 79 | Genetic Defects in Hepatocanalicular Transport. <i>Seminars in Liver Disease</i> , 2000, Volume 20, 365-372.  | 1.8 | 43        |
| 80 | Inherited disorders of transport in the liver. <i>Current Opinion in Genetics and Development</i> , 2000, 10, 310-313.  | 1.5 | 19        |
| 81 | Defects in the enterohepatic circulation of bile acids. <i>Journal of Hepatology</i> , 2000, 32, 5-6.   | 1.8 | 0         |
| 82 | Hepatocanalicular bile salt export pump deficiency in patients with progressive familial intrahepatic cholestasis. <i>Gastroenterology</i> , 1999, 117, 1370-1379.  | 0.6 | 423       |
| 83 | Pediatric liver disease. <i>Current Opinion in Gastroenterology</i> , 1999, 15, 249-252.  | 1.0 | 2         |
| 84 | A gene encoding a liver-specific ABC transporter is mutated in progressive familial intrahepatic cholestasis. <i>Nature Genetics</i> , 1998, 20, 233-238.   | 9.4 | 968       |
| 85 | Gene Structure of the Human Amiloride-Sensitive Epithelial Sodium Channel Beta Subunit. <i>Biochemical and Biophysical Research Communications</i> , 1998, 252, 208-213.  | 1.0 | 29        |
| 86 | Identification of a Locus for Progressive Familial Intrahepatic Cholestasis PFIC2 on Chromosome 2q24. <i>American Journal of Human Genetics</i> , 1997, 61, 630-633.  | 2.6 | 190       |
| 87 | A novel splice site mutation in the $\beta_3$ subunit of the epithelial sodium channel gene in three pseudohypoaldosteronism type 1 families. <i>Nature Genetics</i> , 1996, 13, 248-250.   | 9.4 | 228       |
| 88 | Localisation of Pseudohypoaldosteronism Genes to Chromosome 16p12.2 and 12p13.1-Pter by Homozygosity Mapping. <i>Human Molecular Genetics</i> , 1996, 5, 293-299.   | 1.4 | 73        |