

Table S4

Genes that are up-regulated in the groups of inflammation (in blue) or fibrosis (in red) in children with biliary atresia, presented on descending order based on fold change between the subtypes.

Gene	Description	Fold Change	GO Biological Process Annotation
<i>IL1RL1</i>	Interleukin 1 receptor-like 1	5.617	Immune response Signal transduction
<i>DEFA1</i>	Defensin, alpha 1	5.175	Defense response to bacteria Defense response to fungi Response to virus Xenobiotic metabolism
<i>S100A12</i>	S100 calcium binding protein A12 (calgranulin C)	3.934	Defense response to bacteria Defense response to fungi Inflammatory response Xenobiotic metabolism
<i>HEMGN</i>	Hemogen	3.561	---
<i>HBG1</i>	Hemoglobin, gamma A	3.521	Oxygen transport Transport
<i>RHAG</i>	Rh-associated glycoprotein	3.482	Ammonium transport Circulation Protein complex assembly Transport
<i>HBM</i>	Hemoglobin, mu	3.45	Oxygen transport Transport
<i>S100A8</i>	S100 calcium binding protein A8 (calgranulin A)	3.415	Inflammatory response
<i>CALCA</i>	Calcitonin/calcitonin-related polypeptide, alpha	3.369	G-protein signaling, coupled to cAMP Nucleotide second messenger Adenylate cyclase activation Blood pressure regulation Cell-cell signaling Elevation of cytosolic calcium ion concentration Phospholipase C activation Skeletal development
<i>ZNF165</i>	Zinc finger protein 165	3.298	Regulation of transcription, DNA-dependent

<i>DEFA4</i>	Defensin, alpha 4, corticostatin	3.214	Defense response to bacteria Defense response to fungi Response to pest, pathogen or parasite Xenobiotic metabolism
<i>AKAP12</i>	A kinase (PRKA) anchor protein (gravin) 12	3.132	G-protein coupled receptor signaling Protein targeting Signal transduction
<i>ERAF</i>	Erythroid associated factor	3.13	Hemoglobin metabolism Hemopoiesis Protein folding
<i>GYPA</i>	Glycophorin A (MNS blood group)	2.985	Biological process unknown
<i>HBD</i>	Hemoglobin, delta	2.933	Oxygen transport Transport
<i>CA1</i>	Carbonic anhydrase I	2.922	One-carbon compound metabolism
<i>S100P</i>	S100 calcium binding protein P	2.91	---
<i>SLC4A1</i>	Solute carrier family 4, anion exchanger, member 1	2.874	Anion transport Cell ion homeostasis
<i>MS4A3</i>	Membrane-spanning 4-domains, subfamily A, member 3 (hematopoietic cell-specific)	2.824	Signal transduction
<i>CEACAM8</i>	Carcinoembryonic antigen-related cell adhesion molecule 8	2.7	Immune response
<i>HBA1</i>	Hemoglobin, alpha 1	2.682	---
<i>MAFF</i>	V-maf musculoaponeurotic fibrosarcoma oncogene homolog F (avian)	2.622	Parturition Regulation of transcription, DNA-dependent Transcription from RNA pol II promoter
<i>BRE</i>	Brain and reproductive organ-expressed (TNFRSF1A modulator)	2.617	D-ribose metabolism Carbohydrate metabolism
<i>LTF</i>	Lactotransferrin	2.61	Defense response to bacteria Humoral immune response Iron ion homeostasis Iron ion transport Transport

<i>PROK2</i>	Prokineticin 2	2.562	Activation of MAPK activity Angiogenesis Anti-apoptosis Cell proliferation Chemotaxis Elevation of cytosolic calcium ion conc. Inflammatory response Neuropeptide signaling pathway Smooth muscle contraction Rhythmic process Sensory perception of pain Spermatogenesis
<i>CRISP3</i>	Cysteine-rich secretory protein 3	2.536	Cell-cell adhesion Defense response Fertilization Innate immune response Spermatogenesis
<i>HSPA1B</i>	Heat shock 70kDa protein 1B	2.449	mRNA catabolism Protein folding Response to unfolded protein
<i>SPTA1</i>	Spectrin, alpha, erythrocytic 1 (elliptocytosis 2)	2.395	Actin filament organization Barbed-end actin filament capping Regulation of cell shape
<i>MPO</i>	Myeloperoxidase	2.312	Anti-apoptosis Defense response Hydrogen peroxide catabolism Response to oxidative stress
<i>IL1R2</i>	Interleukin 1 receptor, type II	2.29	Immune response
<i>GYPB</i>	Glycophorin B (MNS blood group)	2.286	---
<i>CLC</i>	Charcot-Leyden crystal protein	2.273	Antimicrobial humoral response (sensu) Vertebrata Development Lipid catabolism Phospholipid metabolism
<i>HBA2</i>	Hemoglobin, alpha 2	2.229	---
<i>RHCE</i>	Rh blood group, CcEe antigens	2.224	Transport
<i>ELA2</i>	Elastase 2, neutrophil	2.201	Proteolysis
<i>PTX3</i>	Pentraxin-related gene, rapidly induced by IL-1 beta	2.188	Inflammatory response

<i>ALAS2</i>	Aminolevulinate, delta-, synthase 2 (sideroblastic/hypochromic anemia)	2.179	Biosynthesis Heme biosynthesis
<i>SLC25A37</i>	Solute carrier family 25, member 37	2.179	Transport
<i>HSPA6</i>	Heat shock 70kDa protein 6 (HSP70B')	2.174	Protein folding Response to unfolded protein
<i>G0S2</i>	G0/G1 switch 2	2.171	Regulation of progression through cell cycle
<i>DNAJB1</i>	DnaJ (Hsp40) homolog, subfamily B, member 1	2.167	Protein folding Response to unfolded protein
<i>XK</i>	X-linked Kx blood group (McLeod syndrome)	2.146	Amino acid transport Transport
<i>C13orf18</i>	Chromosome 13 open reading frame 18	2.145	---
<i>MYB</i>	V-myb myeloblastosis viral oncogene homolog (avian)	2.117	Regulation of transcription Regulation of transcription, DNA-dependent
<i>AFP</i>	Alpha-fetoprotein	2.111	Immune response Transport
<i>RHD</i>	Rh blood group, D antigen	2.096	---
<i>AKR1C2</i>	Aldo-keto reductase family 1, member C2 (dihydrodiol dehydrogenase 2; bile acid binding protein; 3-alpha hydroxysteroid dehydrogenase, type III)	2.09	Xenobiotic metabolism
<i>CDH19</i>	Cadherin 19, type 2	2.072	Homophilic cell adhesion
<i>HSPA1A</i>	Heat shock 70kDa protein 1A	2.037	Protein folding Response to unfolded protein
<i>BPI</i>	Bactericidal/permeability-increasing protein	2.032	Defense response to bacteria Immune response
<i>DNAJA4</i>	DnaJ (Hsp40) homolog, subfamily A, member 4	2.012	Protein folding
<i>MMP9</i>	Matrix metalloproteinase 9 (gelatinase B, 92kDa gelatinase, 92kDa type IV collagenase)	1.996	Collagen catabolism Peptidoglycan metabolism Proteolysis
<i>HBB</i>	Hemoglobin, beta	1.967	Biological process unknown Oxygen transport Transport
<i>SELE</i>	Selectin E (endothelial adhesion molecule 1)	1.905	Cell adhesion Inflammatory response
<i>SNCA</i>	Synuclein, alpha (non A4 component of amyloid precursor)	1.901	Anti-apoptosis Central nervous system development

<i>CTSG</i>	Cathepsin G	1.867	Immune response Proteolysis
<i>PRG2</i>	Proteoglycan 2, bone marrow (NK cell activator, eosinophil granule major basic protein)	1.847	Defense response to bacteria Inflammatory response
<i>EPB42</i>	Erythrocyte membrane protein band 4.2	1.832	Erythrocyte maturation Peptide cross-linking Regulation of cell shape
<i>AGPAT9</i>	Lysophosphatidic acid acyltransferase theta	1.817	Metabolism
<i>ELL2</i>	Elongation factor, RNA polymerase II, 2	1.815	RNA elongation from RNA pol II promoter Regulation of transcription, DNA-dependent
<i>DNTT</i>	Deoxynucleotidyltransferase, terminal	1.805	DNA modification DNA replication Antimicrobial humoral response (sensu) Vertebrata
<i>MMP8</i>	Matrix metalloproteinase 8 (neutrophil collagenase)	1.789	Collagen catabolism Peptidoglycan metabolism Proteolysis
<i>RNASE3</i>	Ribonuclease, RNase A family, 3 (eosinophil cationic protein)	1.744	RNA catabolism Defense response to bacteria
<i>OLFM4</i>	Olfactomedin 4	1.735	---
<i>CHI3L1</i>	Chitinase 3-like 1 (cartilage glycoprotein-39)	1.721	Carbohydrate metabolism Chitin catabolism
<i>TCN1</i>	Transcobalamin I (vitamin B12 binding protein, R binder family)	1.714	Cobalamin transport Cobalt ion transport Ion transport
<i>TRIM55</i>	Tripartite motif-containing 55	1.634	Muscle development Protein ubiquitination Signal transduction
<i>DNAJA1</i>	DnaJ (Hsp40) homolog, subfamily A, member 1	1.593	Protein folding Response to unfolded protein
<i>HSPD1</i>	Heat shock 60kDa protein 1 (chaperonin)	1.563	Cellular protein metabolism Protein folding Protein import into mitochondrial matrix Response to unfolded protein
<i>CAMP</i>	Cathelicidin antimicrobial peptide	1.547	Defense response to bacteria Response to pest, pathogen or parasite

<i>CGA</i>	Glycoprotein hormones, alpha polypeptide	1.511	Cell-cell signaling Signal transduction
<i>FAM129C</i>	B-cell novel protein 1	1.508	---
<i>PIP5K1B</i>	Phosphatidylinositol-4-phosphate 5-kinase, type I, beta	1.475	---
<i>IGSF1</i>	Immunoglobulin superfamily, member 1	1.449	Cell adhesion
<i>IGHM</i>	Immunoglobulin heavy constant mu	1.43	Immune response
<i>ARNTL</i>	Aryl hydrocarbon receptor nuclear translocator-like	1.337	Circadian rhythm Regulation of transcription, DNA-dependent signal transduction
<i>LOC654433</i>	Hypothetical LOC654433	1.018	---
<i>ACSM2B</i>	Acyl-CoA synthetase medium-chain family member 2B	4.288	---
<i>HTR2B</i>	5-hydroxytryptamine (serotonin) receptor 2B	3.934	G-protein signaling, coupled to IP3 second messenger (phospholipase C activating Circulation Regulation of I-kB kinase/NF-kB cascade Signal transduction
<i>COL8A1</i>	Collagen, type VIII, alpha 1	3.249	Cell adhesion Phosphate transport
<i>PTCH1</i>	Patched homolog 1 (Drosophila)	2.682	Cell cycle Cell proliferation Morphogenesis Negative regulation of cell cycle progression Signal transduction
<i>MAP3K13</i>	Mitogen-activated protein kinase kinase kinase 13	2.565	JNK cascade Activation of MAPKK activity Activation of NFkB transcription factor Protein amino acid autophosphorylation
<i>CTHRC1</i>	Collagen triple helix repeat containing 1	2.513	Phosphate transport
<i>SPINK1</i>	Serine peptidase inhibitor, Kazal type 1	2.497	---
<i>PECR</i>	Peroxisomal trans-2-enoyl-CoA reductase	2.366	---

<i>COL11A1</i>	Collagen, type XI, alpha 1	2.259	Cartilage condensation Cell adhesion Cell-cell adhesion Extracellular matrix org. and biogenesis Phosphate transport Sensory perception of sound Visual perception
<i>GOPC</i>	Golgi associated PDZ and coiled-coil motif containing	2.139	ER to Golgi transport Golgi to plasma membrane transport Protein transport
<i>XPO1</i>	Exportin 1 (CRM1 homolog, yeast)	2.139	mRNA export from nucleus mRNA processing Protein import into nucleus, docking Protein transport
<i>HOPX</i>	HOP homeobox	2.121	Development Regulation of transcription, DNA-dependent
<i>FMR1</i>	Fragile X mental retardation 1	2.109	---
<i>TPCN1</i>	Two pore segment channel 1	1.987	---
<i>C8orf70</i>	Chromosome 8 open reading frame 70	1.966	---
<i>FMO2</i>	Flavin containing monooxygenase 2 (non-functional)	1.948	Electron transport
<i>C1orf41</i>	Chromosome 1 open reading frame 41	1.913	---
<i>NHLRC3</i>	Similar to RIKEN cDNA 8030451K01	1.879	Insulin receptor signaling pathway
<i>MLLT3</i>	Myeloid/lymphoid or mixed-lineage leukemia (trithorax homolog, Drosophila); translocated to, 3	1.86	Regulation of transcription, DNA-dependent transcription
<i>ATAD4</i>	ATPase family, AAA domain containing 4	1.853	Protein catabolism
<i>ITPR2</i>	Inositol 1,4,5-triphosphate receptor, type 2	1.842	---
<i>TIA1</i>	TIA1 cytotoxic granule-associated RNA binding protein	1.824	---
<i>SOS1</i>	Son of sevenless homolog 1 (Drosophila)	1.8	Ras protein signal transduction
<i>C17orf42</i>	Chromosome 17 open reading frame 42	1.689	---
<i>CCDC76</i>	Coiled-coil domain containing 76	1.687	---
<i>BCL11B</i>	B-cell CLL/lymphoma 11B (zinc finger protein)	1.636	Regulation of transcription, DNA-dependent

<i>IFI44</i>	Interferon-induced protein 44	1.616	Response to virus
<i>PDE4DIP</i>	Phosphodiesterase 4D interacting protein (myomegalin)	1.616	---
<i>MAP3K1</i>	Mitogen-activated protein kinase kinase kinase 1	1.613	---
<i>LOC389831</i>	Hypothetical gene supported by AL713796	1.597	---
<i>PER3</i>	Period homolog 3 (Drosophila)	1.597	Regulation of transcription, DNA-dependent Signal transduction Transcription
<i>EML4</i>	Echinoderm microtubule associated protein like 4	1.524	---
<i>SFRS18</i>	Chromosome 6 open reading frame 111	1.466	---
<i>TMED10</i>	Transmembrane emp24-like trafficking protein 10 (yeast)	1.465	---
<i>PTP4A1</i>	Protein tyrosine phosphatase type IVA, member 1	1.42	---
<i>ABCA5</i>	ATP-binding cassette, sub-family A (ABC1), member 5	1.406	---
<i>PHACTR2</i>	Phosphatase and actin regulator 2	1.284	Metabolism Nucleobase, nucleoside, nucleotide and nucleic acid metabolism
<i>FARP1</i>	FERM, RhoGEF (ARHGEF) and pleckstrin domain protein 1 (chondrocyte-derived)	1.027	---