

**Opinion paper on the diagnosis and treatment of progressive familial
intrahepatic cholestasis**

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Search #no	Topic	Search Terms
1	Disease	"PFIC" OR "Progressive familial intrahepatic cholestasis" OR "Byler's disease" OR "Byler"
2	Genetic testing	"Genetic testing" OR "Genetic test" OR "Testing" OR "Test" OR "Screening" OR "Screen" OR "Genetic" OR "Gene" OR "Genes" OR "FIC1" OR "PFIC1" OR "BSEP" OR "PFIC2" OR "MDR3" OR "PFIC3" OR "TJP2" OR "PFIC4" OR "FXR" OR "PFIC5" OR "MYO5B" OR "PFIC6" OR "ATP8B1" OR "ABCB11" OR "ABCB4" OR "TJP2" OR "NR1H4" OR "USP53" OR "PFIC7" OR "KIF12" OR "PFIC8"
3	Biomarkers of disease progression	"Serum bile acids" OR "Bile acids" OR "Bile" OR "Bilirubin" OR "Conjugated bilirubin" OR "Biomarker" OR "Biomarkers" OR "Serum biomarker" OR "Diagnosis" OR "Disease progression" OR "Disease progress" OR "Progression" OR "Predict" OR "Prediction" OR "Predictive" OR "Indicate" OR "Indicator"
4	Treatment landscape	"Treatment" OR "Treat" OR "Treating" OR "IBAT inhibitor" OR "Ileal bile acid transport inhibitor" OR "Ileal bile acid transport inhibitors" OR "Therapeutic" OR "Medication" OR "guidelines" OR "guide" OR "Recommend" OR "Recommended" OR "Consensus" OR "Position" OR "Case series" OR "Case" OR "Case reports"
5	Genetic testing in PFIC	1 AND 2
6	Biomarkers of disease progression	1 AND 3
7	PFIC pathway	1 AND 4
8	Date of literature search	03/01/2023

Table S1. Literature search terms and literature search strategy