Supplementary Tables

Outcome after liver transplantation for progressive familial intrahepatic cholestasis: a systematic review and meta-analysis

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Table S1. Overview of studies and patient characteristics – PFIC1

Study (reference)	number of patients	female	alive	genotype	median age last follow-up in months (IQR)	median age of onset in months (IQR)	median age at LT in months (IQR)	LD or DD LT	graft steatosis	diarrhea	SBD post-LT	rejection	re-LT
Lykavieris et al. 2003	2	0%	100%	50% FIC1-B, 50% FIC1-C	180.5 (18.5)	2 (0)	57 (9)	100% DD	100%	100%	NA	NA	0%
Usui et al. 2009	1	0%	100%	100% FIC1-A	160 (0)	12 (0)	60 (0)	100% LD	0%	0%%	NA	NA	100%
Davit-Spraul et al. 2010	6	NA	67%	16% FIC1-A, 34% FIC1-B, 55% FIC1-C	105 (142.5)	2 (1.5)	47.5 (19.75)	100% DD	50% (1 NA)	83%	NA	17%	50%
Cash et al. 2011	1	100%	100%	100% FIC1-C	240 (0)	NA	120 (0)	100% DD	100%	100%	NA	NA	0%
Ebata et al. 2012	1	100%	100%	NA	NA	NA	168 (0)	100% LD	0%	0%	NA	NA	0%
Nicastro et al. 2012	1	0%	100%	100% FIC1-A	NA	5 (0)	43 (0)	100% LD	100%	100%	100%	NA	0%
Berumen et al. 2014	1	100%	100%	NA	108 (0)	NA	84 (0)	100% DD	100%	100%	NA	100%	0%
Cigerli et al. 2014	1	0%	100%	NA	288 (0)	NA	36 (0)	100% LD	0%	0%	NA	NA	NA
Giovannoni et al. 2015	4	NA	NA	25% FIC1-A, 75% FIC1-C	NA	5.5 (3)	NA	100% DD	NA	NA	NA	62%	NA
Mali et al. 2016	1	100%	100%	100% FIC1-B	35 (0)	0.75 (0)	26 (0)	100% DD	0%	0%	NA	NA	NA
Droege et al. 2017	4	0%	100%	50% FIC1-A, 25% FIC1-B, 25% FIC1-C	52 (54.5)	2 (2.25)	37 (32)	NA	NA	NA	NA	NA	25%
Polat et al. 2017	12	NA	100%	NA	100 (0)	NA	50 (0)	NA	NA	NA	NA	NA	NA

Oya et al. 2017	2	50%	50%	100% FIC1-B	192 (0)	NA	90 (54)	100% LD	100%	100%	NA	NA	50%
Alrabadi et al. 2018	2	50%	100%	50% FIC1-A, 50% FIC1-C	119 (49)	NA	55 (29)	100% DD	100%	100%	100%	NA	NA
Bull et al. 2018	24	67%	87.5 %	NA	113 (0)	NA	70 (0)	NA	79%	71%	NA	NA	4%
Liu et al. 2018	3	67%	100%	NA	NA	3 (1)	36 (12)	67% DD	67%	67%	NA	NA	0%
Ruth et al. 2018	6	NA	100%	NA	219 (0)	NA	74 (0)	NA	NA	NA	NA	NA	NA
Stephanie et al. 2018	1	100%	100%	100% FIC1-B	27 (0)	1 (0)	6 (0)	100% LD	100%	100%	NA	NA	0%
Kang et al. 2019	1	100%	100%	100% FIC1-B	53 (0)	1 (0)	26 (0)	NA	100%	100%	NA	0%	0%
Rhee et al. 2019	1	100%	100%	100% FIC1-B	21 (0)	1 (0)	19 (0)	100% DD	100%	100%	NA	NA	0%
Gül-Klein et al. 2021	7	43%	100%	NA	204 (66)	1 (0)	9 (11)	57% DD	42%	NA	NA	NA	0%
Henkel et al. 2021	7	71%	100%	43% FIC1-A, 14% FIC1-C, (3 NA)	212 (0)	2.8 (0)	81 (0)	86% DD	14%	86%	NA	57%	0%
Jankowska et al. 2021 [6](*6*)	1	0%	100%	NA	76 (0)	1.5 (0)	16 (0)	100% LD	100%	100%	100%	NA	0%
Mizutani et al. 2021	3	33%	67%	67% FIC1-A, 33% FIC1-B	144 (90)	6 (2)	36 (54)	100% LD	NA	0%	NA	NA	0%
Okamoto et al. 2021	12	58%	50%	50% FIC1-A, 16% FIC1-B, 34% FIC1-C	212.5 (135.5)	NA	54 (1)	100% LD	75%	83%	NA	8%	25%
Hang et al. 2022	3	33%	100%	33% FIC1-B, 2 NA	204 (25)	NA	84 (25)	67% LD	33%	33%	NA	NA	0%
Hertel et al. 2021	2	NA	100%	NA	228 (12)	2 (0)	114 (42)	NA	NA	NA	NA	NA	NA

Kavallar et al. 2022	3	33%	100%	100% FIC1-C	72 (36)	2 (0.5)	30 (19.5)	67% LD	100%	100%	NA	NA	0%
Masahata et al. 2022	1	0%	0%	NA	142 (0)	3 (0)	136 (0)	100% LD	0%	0%	NA	NA	0%
Mehta et al. 2022	1	0%	0%	100% FIC1-B	54 (0)	1 (0)	54 (0)	100% LD	0%	0%	NA	100%	0%
Namgoong et al. 2022	2	50%	50%	NA	60.5 (25.5)	NA	14 (4)	100% DD	50%	0%	NA	NA	50%
Ohlendorf et al. 2022	1	0%	100%	100% FIC1-C	120 (0)	2 (0)	32 (0)	100% DD	0%	100%	NA	NA	0%
Shanmugam et al. 2022	1	0%	100%	NA	96 (0)	3 (0)	7 (0)	100% LD	100%	100%	100%	NA	NA
Al-Hussaini et al. 2021	2	0%	100%	100% FIC1-C	84 (12)	1 (0)	86 (12)	NA	NA	NA	NA	NA	NA

DD – deceased donor liver transplantation, IQR – interquartile range, LD – living donor liver transplantation, LT – liver transplantation, NA – not available

Table S2. Overview of studies and patient characteristics – PFIC2

Study (Reference)	number of patients	female	alive	genotype	median age last follow-up in months (IQR)	median age of onset in months (IQR)	median age at LT in months (IQR)	LD or DD LT	AIBD	rejection	re-LT
Englert et al. 2007	17	NA	100%	NA	NA	NA	NA	NA	0%	NA	NA
Jara et al. 2009	3	67%	100%	67% BSEP-2, 33% BSEP-3	NA	NA	44 (18)	100% DD	100%	0%	0%
Keitel et al. 2009	1	100%	0%	100% BSEP-2	91 (0)	NA	42 (0)	100% DD	100%	0%	100%
Davit-Spraul et al. 2010	15	NA	87%	13% BSEP-1, 74% BSEP-2, 13% BSEP-3	180 (132)	1 (0)	84 (62)	97% DD	0%	NA	0%
Hori et al. 2010	3	NA	100%	NA	53 (32.5)	NA	17 (24)	100% LD	0%	100%	NA
Maggiore et al. 2010	2	50%	50%	50% BSEP-2, 50% BSEP-3	300 (36)	2.5 (0)	71 (37)	100% DD	100%	50%	0%
Siebold et al. 2010	5	NA	60%	40% BSEP-2, 60% BSEP-3	109 (108)	NA	29 (14)	NA	100%	0%	60%
Shimizu et al. 2011	2	100%	100%	100% BSEP-2	67 (34)	2.5 (0)	13 (4)	100% LD	0%	0%	0%
Lin et al. 2013	2	0%	100%	100% BSEP-3	180 (36)	NA	35 (25)	100% DD	100%	50%	50%
Giovanoni et al. 2015	8	NA	100%	87% BSEP-2, 13% BSEP-3	17 (20.25)	17 (20.25)	NA	NA	0%	NA	NA
Grammatikopoulos et al. 2015	2	50%	100%	100% BSEP-3	24 (74.5)	5.75 (4.25)	26 (4)	NA	100%	100%	50%

Stindt et al. 2015	8	25%	100%	13% BSEP-1, 87% BSEP-3	50.5 (55.5)	NA	31 (0)	NA	88%	25%	NA
Kubitz et al. 2016	1	0%	100%	100% BSEP-3	152 (0)	24 (0)	37 (0)	100% DD	100%	0%	0%
Lemoine et al. 2016	8	NA	NA	NA	31 (0)	NA	NA	NA	NA	NA	NA
Moes et al. 2016	1	0%	100%	NA	66 (0)	NA	36 (0)	NA	100%	NA	0%
Park et al. 2016	2	100%	NA	100% BSEP-2	NA	5 (4)	72 (48)	NA	NA	NA	NA
Shagrani et al. 2016	2	50%	100%	100% BSEP-3	198 (30)	NA	150 (30)	100% DD	100%	NA	0%
Droege et al. 2017	13	62%	100%	38% BSEP-1, 38% BSEP-2, 24% BSEP-3	24 (45)	6 (9.5)	18 (36)	NA	31%	NA	NA
Patel et al. 2017	1	100%	100%	100% BSEP-3	167 (0)	5 (0)	18 (0)	100% DD	100%	100%	0%
Polat et al. 2017	38	NA	100%	NA	100 (0)	NA	50 (0)	NA	0%	NA	0%
Prusinskas et al. 2017	1	100%	100%	100% BSEP-3	185 (0)	2 (0)	75 (0)	NA	100%	0%	100%
Brinkert et al. 2018	1	0%	100%	100% BSEP-3	96 (0)	10 (0)	16 (0)	100% LD	100%	0%	100%
Bull et al. 2018	33	52%	100%	55% BSEP-1 (15 NA)	80 (0)	NA	37 (0)	NA	0%	67%	NA
Liu et al. 2018	1	0%	100%	NA	79 (0)	NA	60 (0)	100% DD	0%	NA	NA
Ruth et al. 2018	7	NA	100%	NA	181 (0)	NA	62 (0)	NA	0%	NA	0%
Kang et al. 2019	3	100%	NA	33% BSEP-3 (2 NA)	56 (44)	1 (0)	NA	NA	33%	NA	NA
Quintero et al. 2020	1	100%	100%	100% BSEP-3	100 (0)	6 (0)	14 (0)	NA	100%	100%	0%

Hertel et al. 2021	2	NA	100%	NA	120 (0)	4 (0)	36 (0)	NA	0%	NA	0%
Namgoong et al. 2021	1	100%	100%	NA	96 (0)	7 (0)	7 (0)	100% LD	0%	NA	NA
Akhtar et al. 2022	2	50% (1 NA)	100%	50% BSEP-2, 50% BSEP-3	NA	NA	NA	50% LD (1 NA)	100%	NA	0%
Hang et al. 2022	24	25%	96%	25% BSEP-2, 4% BSEP-3, (17 NA)	60 (0)	NA	11 (20.75)	79% LD	0%	NA	8%
Masahata et al. 2022	2	NA	50%	50% BSEP-3 (1 NA)	162.5 (99.5)	NA	40.5 (17.5)	100% LD	50%	NA	NA
Mehta et al. 2022	1	NA	100%	100% BSEP-2	20 (0)	NA	19 (0)	100% LD	0%	NA	NA
Romeres et al. 2022	1	0%	100%	100% BSEP-3	63 (0)	1 (0)	21 (0)	NA	100%	0%	0%
Al-Hussaini et al. 2021	16	50%	100%	44% BSEP-2, 56% BSEP-3	32 (25.5)	1 (1)	30 (25.5)	NA	NA	NA	NA
Pfister et al. 2023	22	NA	100%	41% BSEP-1, 18% BSEP-2, 41% BSEP-3	40.5 (67.5)	NA	38.5 (67.5)	NA	4.5%	NA	9%

DD – deceased donor liver transplantation, IQR – interquartile range, LD – living donor liver transplantation, LT – liver transplantation, NA – not available

Table S3. Overview of studies and patient characteristics – PFIC3

Study (Reference)	number of patients	female	alive	genotype	median age last follow-up in months (IQR)	median age of onset in months (IQR)	median age at LT in months (IQR)	LD or DD LT	rejection	re-LT
De Vree et al. 1998	2	0	100%	100% MDR3-3	36 (0)	5.5 (2.5)	75 (33)	NA	NA	0% (1 NA)
Jacquemin et al. 2001	16	71% (9 NA)	100%	47% MDR3-1, 31% MDR3-3, (4 NA)	66 (60)	12 (15)	65 (60)	NA	NA	0%
Keitel et al. 2005	3	NA	100%	100% MDR3-1	94 (40.5)	6 (1)	93 (40.5)	NA	NA	NA
Englert et al. 2007	6	NA	100%	NA	NA	NA	NA	NA	NA	NA
Anheim et al. 2010	1	0	100%	100% MDR3-2	NA	420 (0)	528 (0)	NA	NA	NA
Colombo et al. 2011	5	20%	100%	40% MDR3-1, 40% MDR3-2, 20% MDR3-3	12 (12)	24 (17)	82 (39)	NA	NA	0%
Kaur et al. 2012	2	0%	100%	NA	45 (11)	6.5 (2.5)	NA	100% LD	NA	0%
Giovanoni et al. 2015	2	0 (1 NA)	100%	100% MDR3-3	24 (0)	33.5 (14.5)	84 (0)	NA	NA	0%
Gordo-Gilart et al. 2015	2	100%	100%	50% MDR3-1, 50% MDR3-3	NA	16.5 (1.5)	73.2 (13.2)	NA	NA	NA
Vij et al. 2015	3	67%	100%	NA	10 (15.5)	18 (6)	72 (18)	66.7% (1 NA)	NA	0%
Agarwal et al. 2016	2	0%	100%	NA	5.5 (3.5)	NA	NA	50% LD	NA	0%
Xiang et al. 2017	1	0%	100%	100% MDR3-1	6 (0)	216 (0)	216 (0)	100% DD	NA	0%
Droege et al. 2017	5	40%	100%	80% MDR3-1, 20% MDR3-2	96 (84)	7.5 (10)	48 (24)	NA	NA	60%
Liu et al. 2018	1	NA	83.3%	NA	12 (0)	NA	84 (0)	100% DD	NA	0%

Schatz et al. 2018	13	38%	84.6%	85% MDR3-1, 15% MDR3-3	70 (112)	25 (42)	70 (112)	NA	NA	0%
Acar et al. 2019	13	23%	100%	NA	65 (0)	NA	29 (0)	100% LD	NA	0%
Goubran et al. 2020	1	100%	100%	100% MDR3-1	216 (0)	18 (0)	204 (0)	NA	100%	0%
Hertel et al. 2021	1	NA	100%	NA	156 (0)	15 (0)	48 (0)	NA	NA	0%
Lipinski et al. 2020	2	50%	100%	100% MDR3-1	54 (30)	51 (21)	186 (42)	50% DD (1 NA)	NA	0%
Hang et al. 2022	12	25%	100%	8% MDR3-1, 16% MDR3-2, (9 NA)	60 (48)	NA	89 (61.25)	75% LD	NA	0%
Masahata et al. 2022	1	0%	100%	NA	8 (0)	2 (0)	72 (0)	100% LD	100%	0%
Sinha et al. 2022	3	100%	100%	67% MDR3-2, 33% MDR3-3	84 (0)	216 (204)	528 (123)	100% DD	NA	0% (2 NA)
Zhu et al. 2022	1	0%	100%	100% MDR3-1	18 (0)	192 (0)	204 (0)	100% DD	NA	0%
Al-Hussaini et al. 2021	7	57%	100%	43% MDR3-1, 57% MDR3-3	110 (14)	10 (8.5)	108 (14)	NA	NA	NA

DD – deceased donor liver transplantation, IQR – interquartile range, LD – living donor liver transplantation, LT – liver transplantation, NA – not available

Table S4. Overview of studies and patient characteristics – PFIC4

Study (reference)	number of patients	female	alive	median age last follow-up in months (IQR)	median age of onset in months (IQR)	median age at LT in months (IQR)	LD or DD LT	rejection	re-LT
Sambrotta et al. 2013	9	NA	100%	49 (36)	2 (1)	48 (30)	NA	NA	NA
Zhou et al. 2015	1	0	100%	25 (0)	2 (0)	24 (0)	NA	NA	NA
Alvarez et al. 2017	2	100%	100%	65.5 (40.5)	1 (0)	17.5 (4.5)	50% LD	NA	NA
Vij et al. 2017	1	100%	100%	85 (0)	3 (0)	84 (0)	DD	NA	NA
Wei et al. 2020	2	50%	100%	355 (78)	330 (102)	354 (78)	100% DD	NA	NA
Menon et al. 2021	2	50%	100%	73 (24)	1 (0)	72 (24)	50% LD	NA	NA
Kumar et al. 2022	9	NA	100%	10 (11)	NA	9 (10)	NA	NA	NA
Masahata et al. 2022	1	100%	100%	108 (0)	1 (0)	48 (0)	NA	NA	NA
Mehta et al. 2022	2	50%	100%	26 (6)	NA	25 (6)	100% LD	NA	NA

DD – deceased donor liver transplantation, IQR – interquartile range, LD – living donor liver transplantation, LT – liver transplantation, NA – not available

Table S5. Overview risk of bias assesment

Study	PFIC	Type of study	n	Item 1	Item 2	Item 3	Item 4	Item 5	Item 6	Item 7	Item 8	Item 9	Item 10	Item 11	sum	max	ROB
Alrabadi 2018	PFIC 1	Case Report	2	1	1	1	1	1	1	1	1				8	8	**low**
Berumen 2014	PFIC 1	Case Report	1	0	1	0	1	1	1	1	1				6	8	**moderate**
Bull 2018	PFIC 1/2	Cohort Study	42/60	1	1	1	1	1	0	1	0	1	1	1	9	11	**low**
Cash 2011	PFIC 1	Case Report	1	1	1	1	1	1	1	NA	1				7	7	**low**
Cigerli 2014	PFIC 1	Case Report	1	1	1	1	1	1	0	NA	1				6	7	**low**
Davit-Spraul 2010	PFIC 1/2	Cohort Study	13/39	1	1	1	0	0	1	1	1	1	0	1	8	11	**moderate**
Droege 2017	PFIC 1/2/3	Cohort Study	18/88/43	1	1	1	0	0	NA	NA	NA	NA	NA	1	4	6	**moderate**
Ebata 2012	PFIC 1	Case Report	1	0	1	1	1	1	1	NA	1				6	7	**low**
Giovannoni 2015	PFIC 1/2/3	Case Series	7/17/3	0	1	1	1	0	0	1	1	0	NA		5	9	**serious**
Gül-Klein 2021	PFIC 1	Case Series	7	1	0	1	1	1	1	1	1	0	1		8	10	**moderate**
Hang 2022	PFIC 1/2/3	Cohort Study	4/25/12	1	1	1	0	0	1	1	1	0	0	1	7	11	**moderate**
Henkel 2021	PFIC 1	Case Series	7	1	1	1	1	1	1	1	1	0	1		9	10	**low**
Hertel 2021	PFIC 1/2/3	Cohort Study	26/53/19	1	1	1	0	0	NA	NA	NA	NA	NA	1	4	6	**moderate**
Jankowska 2021	PFIC 1	Case Report	2	1	1	1	0	1	1	1	1				7	8	**low**

Kang 2019	PFIC 1/2	Case Series	1/4	1	1	1	1	1	0	1	1	0	NA		7	9	**moderate**
Kavallar 2022	PFIC 1	Case Report	3	0	1	1	1	1	1	1	1				7	8	**low**
Liu 2018	PFIC 1/2/3	Case Series	3/1/1	0	1	1	1	1	0	1	1	0	1		7	10	**moderate**
Lykavieris 2003	PFIC 1	Case Report	2	0	1	1	1	1	1	1	1				7	8	**low**
Mali 2016	PFIC 1	Case Report	1	1	0	1	1	1	1	1	1				7	8	**low**
Masahata 2022	PFIC 1	Case Series	6	0	1	1	1	0	0	1	1	0	NA		5	9	**serious**
Mehta 2022	PFIC 1/2/3/4	Case Series	4/3/2/2	0	1	1	1	0	0	1	1	0	NA		5	9	**serious**
Mizutani 2021	PFIC 1/BRIC1	Case Series	10/5	0	1	1	0	0	1	0	NA	1	1		5	9	**serious**
Nicastro 2012	PFIC 1	Case Report	1	1	0	1	1	1	1	1	1				7	8	**low**
Ohlendorf 2022	PFIC 1	Case Report	1	1	1	1	1	1	1	1	1				8	8	**low**
Okamoto 2021	PFIC 1	Case Series	12	0	1	1	1	1	0	1	1	0	1		7	10	**moderate**
Oya 2017	PFIC 1	Case Report	2	0	0	0	1	1	1	1	1				5	8	**moderate**
Polat 2017	PFIC 1/2/3	Abstract ESPGHAN	12/38/11	0	0	0	1	1	1	0	1	1	1	1	7	11	**moderate**
Rhee 2019	PFIC 1	Case Report	1	0	1	1	1	0	1	NA	1				5	7	**moderate**
Ruth 2018	PFIC 1/2/3	Abstract ESPGHAN	8/20/2	0	1	1	1	1	1	0	0	1	1	1	8	11	**moderate**
Shanmugam 2022	PFIC 1	Case Report	1	0	0	1	1	1	1	1	1				6	8	**moderate**
Stephanie 2018	PFIC 1	Case Report	1	0	1	0	1	1	1	NA	1				5	7	**moderate**

Usui 2009	PFIC 1	Case Report	1	0	1	1	1	1	1	NA	1				6	7	**low**
Akhtar 2022	PFIC 2	Abstract ESPGHAN	4	0	0	0	1	0	NA	0	NA	NA	1	1	3	8	**serious**
Brinkert 2018	PFIC 2	Case Report	1	0	1	1	1	1	1	1	1				7	8	**low**
Englert 2007	PFIC 2/3	Cohort Study	26/16	1	1	1	NA	NA	NA	0	0	0	0	1	4	8	**serious**
Grammatikopoulos 2015	PFIC 2	Case Report	2	0	1	1	1	1	1	1	1				7	8	**low**
Hori 2010	PFIC 1/2	Case Series	11/3	1	0	0	1	1	0	1	1	0	1		6	10	**moderate**
Jara 2009	PFIC 2	Case Report	3	0	1	1	1	1	1	1	1				7	8	**low**
Keitel 2009	PFIC 2	Case Report	1	0	1	0	1	1	1	1	1				6	8	**moderate**
Kubitz 2014	PFIC 2	Case Series	12	0	1	1	0	0	0	0	1	0	NA		3	9	**serious**
Lemoine 2016	PFIC 1/2/3	Cohort Study	10/13/1	1	1	1	0	0	NA	1	1	0	0	1	6	10	**moderate**
Lin 2013	PFIC 2	Case Report	2	0	1	1	1	1	1	1	1				7	8	**low**
Maggiore 2010	PFIC 2	Case Report	2	0	1	1	1	1	1	1	1				7	8	**low**
Masahata 2016	PFIC 2	Case report	1 //12	0	1	1	1	1	1	1	1				7	8	**low**
Moes 2016	PFIC 2	Abstract ESPGHAN	1	0	0	0	1	1	NA	0	NA	NA	1	1	4	8	**serious**
Namgoong 2021	PFIC 2	Case Report	1	0	1	1	0	1	1	NA	1				5	7	**moderate**
Park 2016	PFIC 2	Case Series	5	0	1	1	1	1	0	1	0	1	NA		6	9	**moderate**
Patel 2017	PFIC 2	Case Report	1	0	1	1	1	1	1	1	1				7	8	**low**
Prusinskas 2017	PFIC 2	Case Report	1	0	1	1	1	1	1	1	1				7	8	**low**

Quintero 2020	PFIC 2	Case Report	1	0	1	1	1	1	1	1	1				7	8	**low**
Romeres 2022	PFIC 2	Case Report	1	0	1	1	1	1	1	NA	1				6	7	**low**
Shagrani 2016	PFIC 2	Abstract ESPGHAN	2	0	0	1	1	1	NA	0	NA	NA	1	1	5	8	**moderate**
Shimizu 2011	PFIC 2	Case Report	2	0	1	1	1	1	1	0	1				6	8	**moderate**
Siebold 2010	PFIC 2	Case Series	6	0	1	1	0	0	0	1	1	0	NA		4	9	**serious**
Stindt 2015	PFIC 2	Case Series	7	0	1	1	0	0	1	1	1	0	1		6	10	**moderate**
Acar 2019	PFIC 3	Abstract ESPGHAN	22	0	1	0	1	1	1	0	NA	1	1	1	7	10	**moderate**
Agarwal 2016	PFIC 1/2/3	Cohort Study	2/19/3	1	1	1	1	0	1	1	1	0	0	NA	7	10	**moderate**
Anheim 2010	PFIC 3	Case Report/Letter to the Editor	1	1	0	1	1	1	0	NA	1				5	7	**moderate**
Colombo 2011	PFIC 3	Case Series	28	1	1	1	0	1	1	1	1	1	1		9	10	**low**
De Vree 1998	PFIC 3	Case Report	2	1	1	1	1	0	0	NA	1				5	7	**moderate**
Giovanoni 2011	PFIC 3	Case Report	2	1	1	1	1	NA	NA	NA	1				5	5	**low**
Gordo-Gilart 2015	PFIC 3	Case Series	6	0	1	1	0	0	0	1	NA	0	1		4	9	**serious**
Goubran 2020	PFIC 3	Case Report	1	1	1	1	1	1	1	1	1				8	8	**low**
Jacquemin 2001	PFIC 3	Case Series	31	1	0	1	0	0	1	1	1	1	1		7	10	**moderate**
Kaur 2012	PFIC 1/2/3	Case Series	3/2/2	1	0	1	1	0	0	1	1	0	NA		5	9	**serious**
Keitel 2005	PFIC 2/3	Case Series	5/5	0	1	1	1	0	1	NA	NA	0	1		5	8	**moderate**

Lipinski 2021	PFIC 3	Case Series	4	0	1	1	0	0	0	1	1	0	NA		4	9	**serious**
Schatz 2018	PFIC 3	Cohort Study	38	1	1	1	1	1	NA	1	1	0	0	1	8	10	**moderate**
Sinha 2022	PFIC 3	Case Series	7	0	0	1	0	0	0	1	1	0	NA		3	9	**serious**
Vij 2015	PFIC 3	Case Series	10	1	1	1	1	1	0	1	1	0	NA		7	9	**moderate**
Xiang 2017	PFIC 3	Case Report	1	1	1	1	1	1	1	NA	1				7	7	**low**
Zhu 2022	PFIC 3	Case Report	1	1	1	1	1	1	1	NA	1				7	7	**low**
Alvarez 2017	PFIC 4	Abstract ESPGHAN	3	0	0	1	1	1	NA	1	NA	NA	1	1	6	8	**moderate**
Kumar 2022	PFIC 4	Abstract ESPGHAN	9	0	0	0	1	1	0	0	0	1	1	1	5	10	**serious**
Menon 2021	PFIC 4	Abstract ESPGHAN	3	0	0	0	1	1	NA	0	NA	NA	1	1	4	11	**serious**
Sambrotta 2013	PFIC 4	Case Series	12	0	1	1	0	0	0	1	NA	0	NA		3	8	**serious**
Vij 2017	PFIC 4	Case Report	1	0	1	1	0	0	1	NA	1				4	7	**serious**
Wei 2020	PFIC 4	Case Report	1	1	1	1	1	1	0	NA	1				6	7	**low**
Zhou 2015	PFIC 4	Case Report	2	1	0	1	1	0	0	0	1				4	8	**serious**

NA – not available, ROB – risk of bias