



Dr Palak Trivedi
The UK PSC Study

Recruitment centre: XXXX

Research Centre:
Dept. of Medical Genetics
Lv 6, Addenbrooke's Treatment Centre
Cambridge Biomedical Campus
Hills Rd
Cambridge
CB2 0QQ

***Research study into the genetic causes of Primary
Sclerosing Cholangitis and childhood autoimmune liver disease***
Clinical Data Update – version 1

Question 1

1.1) Is the patient still alive? Yes ☐ No ☐

If yes, please go to Q.2.

1.2) What is the cause of death documented on the death certificate or in the hospital notes (if known)?

Ia.....

Ib.....

Ic.....

1.3) Please also code the cause of death

Liver failure ☐ Hepatobiliary cancer ☐

Colon cancer ☐ Other cancer ☐

Post-Liver Transplant Complications ☐ Sepsis ☐

Other cause ☐ Unknown ☐

1.4.) What was the date of death? (DD/MM/YYYY)

Question 2

2.1) Has the patient had a liver transplant? Yes ☐ No ☐

2.2) What was the indication for the liver transplant? Please tick below

Indication	Tick if applicable
End-stage liver disease with synthetic failure	
Recurrent cholangitis	
Complications of portal hypertension	
Quality of Life issues (itching/fatigue)	
CCA	
HCC	
Recurrent disease	
Biliary dysplasia	

2.3) What was the patient MELD/PELD at the time of liver transplantation?

MELD/PELD Date(DD/MM/YYYY)
(please delete as appropriate)

Unknown MELD/PELD at Transplant ☐

Please use the websites below to calculate the MELD or PELD score (according to the patient's age)

<https://www.mdcalc.com/meld-score-model-end-stage-liver-disease-12-older>

<https://www.mdcalc.com/peld-score-pediatric-end-stage-liver-disease-younger-12>

2.4) Is the patient on the liver transplant waiting list? Yes ☐ No ☐

If no, move straight to Question 3.

2.5) Date the patient was **listed** for liver transplant (if known): _____(DD/MM/YYYY)

2.6) Date of actual liver transplant (if known): _____(DD/MM/YYYY)

2.7) Name of transplant centre (if known) _____

2.8) What were the following blood test results just prior to transplantation?

	RESULT	REFERENCE RANGE	DATE OF TEST
TOTAL BILIRUBIN			
AST			
ALT			
ALP			
GGT			
ALBUMIN			
CREATININE			
SODIUM			
HAEMOGLOBIN			
PLATELETS			
WHITE CELL COUNT			
EOSINOPHIL COUNT			
PROTHROMBIN TIME			
APTT			
INR			

Please document Upper limit of normal for total bilirubin, AST, ALT, ALP and GGT in Reference range column. Please document Lower limit of normal for Albumin and Platelets in Reference Range column

2.9) Does the patient have recurrent PSC or childhood AILD post-transplant?

Yes ☐

No ☐

2.10) Date of diagnosis of recurrent PSC or childhood AILD: _____(DD/MM/YYYY)

Question 3

3.1) Please provide the results of the patient's **most recent** blood tests

****IF THE PATIENT IS A TRANSPLANT RECIPIENT, THEN PLEASE PROVIDE BLOOD TEST RESULTS IMMEDIATELY PRIOR TO TRANSPLANTATION****

	RESULT	REFERENCE RANGE	DATE OF TEST
TOTAL BILIRUBIN			
AST			
ALT			
ALP			
GGT			
ALBUMIN			
CREATININE			
SODIUM			
HAEMOGLOBIN			
PLATELETS			
WHITE CELL COUNT			
EOSINOPHIL COUNT			
PROTHROMBIN TIME			
APTT			
INR			

Please document Upper limit of normal for total bilirubin, AST, ALT, ALP and GGT in Reference range column.

Please document Lower limit of normal for Albumin and Platelets in Reference Range column

Question 4

4.1) Please provide the date of the patient's **most recent** MRCP or ERCP: _____ (DD/MM/YYYY)

4.2) Please indicate any changes seen on **most recent** MRCP or ERCP:

	YES	NO	UNCLEAR
INTRAHEPATIC CHOLANGIOGRAPHIC CHANGES			
EXTRAHEPATIC CHOLANGIOGRAPHIC CHANGES			
BOTH INTRA AND EXTRAHEPATIC CHANGES			
NORMAL (NO EVIDENCE OF CHOLANGIOPATHY)			

4.3) Are digital or DICOM copies of ERCP/MRCP images available?

Yes

☐

No

☐

Question 5

5.1) Does the patient have Inflammatory Bowel Disease?

Yes

☐

No

☐

If no, please proceed to Q. 5.6

5.2) If yes to 5.1, please indicate which type:

	YES	No
ULCERATIVE COLITIS		
INDETERMINATE COLITIS		
CROHN'S COLITIS		

5.3) Date of diagnosis of IBD (if known) _____ (DD/MM/YYYY)

5.4) If they have colitis is it:

Macroscopic

☐

Microscopic

☐

5.5) Distribution of the Colitis (Please tick all that apply)

TERMINAL ILEUM		CAECUM		ASCENDING COLON	
TRANSVERSE COLON		DESCENDING COLON		SIGMOID COLON	
RECTUM					

5.6) Has the patient has ever been investigated for dysplasia? Yes

☐

No

☐

If no, please go to question 5.10

5.7) If the patient has been investigated for dysplasia, what were the findings?

Negative for dysplasia ☐ Indefinite for dysplasia ☐ Low grade dysplasia ☐
 High grade dysplasia ☐ Intramucosal adenocarcinoma ☐
 Invasive adenocarcinoma ☐

5.8) What was the endoscopic appearance of the dysplasia?

Visible, polypoid ☐ Visible, non-polypoid ☐ Invisible ☐

5.9) On what date was the dysplasia detected? _____ (DD/MM/YYYY)

5.10) Has the patient had a colectomy? Yes ☐ No ☐

5.11) If yes, what was the date of their colectomy (if known)? _____ (DD/MM/YYYY)

5.12) Specify type of colectomy (if known): Sub-total colectomy with ileo-anal pouch ☐
 Pan-proctocolectomy with ileostomy ☐

5.13) What was the indication for the colectomy?

Active disease ☐ Active disease and Neoplasia ☐ Low grade dysplasia ☐
 High grade dysplasia ☐ Adenocarcinoma ☐ Other ☐
 Unknown ☐

5.14) If patient has had colon cancer, please list the site: _____

5.15) Has the patient received any biologic treatments for colitis?

Yes ☐ No ☐

5.16) If yes, which treatments were they?

Anti-TNF ☐ Vedolizumab ☐
 Other ☐ Please provide treatment name _____

Question 6

6.1) Has the patient had any of the following:

CONDITION	YES	DATE OF FIRST DIAGNOSIS
CHOLANGIOCARCINOMA		
GALLBLADDER CANCER		
HEPATOCELLULAR CARCINOMA		

PANCREATIC CANCER		
CHOLECYSTECTOMY		
GALLBLADDER DYSPLASIA (ON CHOLECYSTECTOMY REPORT)		
COLORECTAL CANCER		
BILE DUCT DYSPLASIA (FROM ERCP OR SURGERY)		
UNCLASSIFIED HEPATOBILIARY CANCER		

Question 7

7.1) Please put the details of the **most recent** US the patient had (DD/MM/YYYY)

Date of ultrasound

Is liver heterogenous? Yes ☐ No ☐ Not sure ☐

Is capsule of liver irregular? Yes ☐ No ☐ Not sure ☐

Size of spleen (cm).....

If no size recorded, was the spleen: Normal sized ☐ Enlarged ☐

Gallstones? Yes ☐ No ☐

Gallbladder polyps? Yes ☐ No ☐

Ascites? Yes ☐ No ☐

Bile duct dilatation Yes ☐ No ☐

Lymph nodes at the porta? Yes ☐ No ☐

Question 8

8.1) Has the patient had any of the following complications/diagnoses?

	YES	DATE OF FIRST EPISODE OR FIRST NOTED
VARICES ON SCREENING ENDOSCOPY		
VARICEAL BLEED		
ASCITES		
ENCEPHALOPATHY		
CHOLANGITIS (BILIARY SEPSIS)		
HEPATORENAL SYNDROME		
JAUNDICE		
CIRRHOSIS		

Question 9

9.1) Has the patient had a Fibroscan? Yes ☐ No ☐

9.2) If yes, please provide the following information:

Fibroscan date (DD/MM/YYYY)	
Probe type	
Median elastography (kPa)	
IQR Elastography	
Success rate (%)	
CAP	
IQR CAP	

No more questions. Thank you for completing the questionnaire.

FINAL CHECKLIST

Have you included an anonymised copy of the following, stating the patient's UK PSC Study number?

ERCP Report

☐

MRCP Report

☐

Liver Biopsy Report

☐

*If you have any **queries** regarding the **completion** of this questionnaire please contact the UK PSC team (ukpsc@uhb.nhs.uk; Tel: 0121 371 8101).*

Please return completed questionnaires to:
The UK PSC Study,
 Box 238,
 Dept. of Medical Genetics
 Lv 6 Addenbrooke's Treatment Centre,
 Hills Rd,
 Cambridge, CB2 0QQ