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The UK PSC Study

Recruitment centre: XXXX

Research Centre:
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Research study into the genetic causes of Primary Sclerosing Cholangitis and childhood autoimmune liver disease

Paediatric Clinician Questionnaire, version 1

Question 1

1.1) What is the patient's date of birth?..... (DD/MM/YYYY)

1.2) What is the patient's gender? Male ☐ Female ☐

1.3) What is the patient's NHS no. or CHI no.?

1.4) Is the patient under follow-up at your hospital? Yes ☐ No ☐

1.5) What is the name of your hospital?

1.6) Was the patient under another NHS Trust before you started looking after them?

Yes ☐ No ☐

1.7) If the answer to Q1.6 was yes, please name the trust and hospital that was looking after the patient

.....

Question 2

2.1) Is the patient known to have childhood autoimmune liver disease (AILD)?

Yes ☐ No ☐

2.2) Date of Diagnosis of AILD(DD/MM/YYYY)
(i.e confirmation on biopsy or cholangiography, if known)

2.3) Date of first abnormal cholangiogram(DD/MM/YYYY)

2.4) Childhood AILD phenotype

Large-Duct PSC ☐ Small-Duct PSC ☐ Unclassified sclerosing cholangitis ☐
PSC with Features of AIH (PSC/AIH Overlap) ☐ IgG4 associated cholangitis ☐
AIH without PSC ☐ Unknown ☐

2.5) Was the patient initially **diagnosed** with another liver condition?

Yes ☐ No ☐

If no, please go to question 2.8

2.6) If yes to Q2.5, what was the name of this condition?

2.7) On what date was this diagnosis made?..... (DD/MM/YYYY)

2.8) Has the patient had any pre-transplant hepatobiliary surgery? Yes ☐ No ☐

Date of Surgery _____(DD/MM/YYYY) Surgery name _____

Findings: **Please attach a copy of any surgery reports, indicating the patient UK PSC study ID.**

Question 3

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

If yes, please go to Q.4.

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

1a.....

1b.....

1c.....

3.3) Please also code the cause of death

Liver failure ☐ Hepatobiliary cancer ☐ Colon cancer ☐
Other cancer ☐ Post-Liver Transplant Complications ☐ Sepsis ☐
Other cause ☐ Unknown ☐

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

Question 4

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

4.2) What was the indication for the liver transplant?

End-stage liver disease with synthetic failure	<input type="checkbox"/>	Recurrent cholangitis	<input type="checkbox"/>
Complications of portal hypertension	<input type="checkbox"/>	QoL (itching/fatigue)	<input type="checkbox"/>
CCA	<input type="checkbox"/>	HCC	<input type="checkbox"/>
Recurrent disease	<input type="checkbox"/>	Biliary dysplasia	<input type="checkbox"/>

4.3) 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>

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

If no, move straight to Question 5.

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

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

4.7) Name of transplant centre (if known)

4.8) Does the patient have recurrent autoimmune liver disease post-transplant?

Yes ☐ No ☐

4.9) Date of diagnosis of recurrent autoimmune liver disease: (DD/MM/YYYY)

Question 5

5.1) Please provide the blood results **at the time of the diagnosis**

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

5.2) Please provide the results of the patient's blood tests **one year after initial diagnosis**

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

5.3) Please provide the blood results **two years after initial diagnosis**

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

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

****If patient is a transplant recipient, please provide test results immediately pre OLT****

	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 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 6

6.1) Please provide the following immunology results (if multiple results available, please state the earliest result, post-diagnosis)

	RESULT	REF RANGE	TITRE	DATE OF TEST
P-ANCA (POSITIVE OR NEGATIVE)		N/A		
ANA (POSITIVE OR NEGATIVE)		N/A		
SMA [*] (POSITIVE OR NEGATIVE) [*] SMOOTH MUSCLE ANTIBODY	RESULT:	N/A		
ANTI MITOCHONDRIAL ANTIBODY (POSITIVE OR NEGATIVE)		N/A		
LKM-1 ANTIBODY (POSITIVE OR NEGATIVE)		N/A		
IMMUNOGLOBULIN IGG (LEVEL)				
IMMUNOGLOBULIN IGA (LEVEL)				
IMMUNOGLOBULIN IGM (LEVEL)				
IGG4 (LEVEL AND UNITS)				
CA19-9 (LEVEL)				

LC-1				
ANTI SLA				

Please document Upper limit of normal for Immunoglobulin IGG and IgG4 levels in the Reference range column.

Question 7

7.1) Please indicate which of the following was used as the **initial diagnostic** investigation:

INVESTIGATION	YES	NO	DATE
MRCP			
ERCP			
LIVER BIOPSY			

7.2) Was ERCP or MRCP compatible with a cholangiopathy?

Yes ☐

No ☐

7.3) If the patient has had an ERCP or MRCP, did it show:

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

(Please note that extrahepatic biliary tree involves up to the point of bifurcation of the left and right hepatic ducts i.e. everything up to, and including the common hepatic duct).

7.4) Has the patient had a **follow-up** investigation? MRCP ☐ ERCP ☐

7.5) If yes to Q7.4 , please provide the date:..... (DD/MM/YYYY)

7.6) Please indicate 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)			

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

Yes ☐

No ☐

7.8) If the patient has had a Liver Biopsy, please indicate the following findings:

FINDINGS	YES	DATE
LIVER HISTOLOGY COMPATIBLE WITH PSC		
LIVER HISTOLOGY NON-DIAGNOSTIC		
LIVER HISTOLOGY NOT COMPATIBLE WITH PSC		
LIVER BIOPSY NOT UNDERTAKEN		NA
LIVER HISTOLOGY COMPATIBLE WITH AIH		
LIVER HISTOLOGY COMPATIBLE WITH AIH WITH CHOLANGIOPATHY		
LIVER HISTOLOGY COMPATIBLE WITH SCLEROSING CHOLANGITIS		
LIVER HISTOLOGY NOT COMPATIBLE WITH AIH		

7.9) Which stage of liver disease does the patient have (if known)? (Stage 1-4)

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

7.11) If yes, please provide the following information:

FIBROSCAN DATE (DD/MM/YYYY)	
PROBE TYPE	
MEDIAN ELASTOGRAPHY (kPa)	
IQR ELASTOGRAPHY	
SUCCESS RATE (%)	
CAP	
IQR CAP	

7.12) Which of the following investigations have been undertaken?

INVESTIGATION	YES/NO/ UNKNOWN	DATE	RESULT	COMMENTS
BILIARY DILATION				
BILIARY STENT				
BRUSHING/BIOPSY				
CHOLANGIOSCOPY				
GASTROSCOPY				
COLONOSCOPY (TO SCREEN FOR IBD)				
CT				
MRI				
MRE				

****PLEASE INCLUDE AN ANONYMISED COPY OF THE REPORTS CITED ABOVE, STATING THE PATIENT UK-PSC STUDY ID****

Question 8

8.1) Does the patient have Inflammatory Bowel Disease? Yes ☐ No ☐

If no, please proceed to Q. 8.6

8.2) If yes to 8.1, please indicate which type:

	YES	NO
ULCERATIVE COLITIS		
INDETERMINATE COLITIS		
CROHNS COLITIS		

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

8.4) If they have colitis is it: MACROSCOPIC ☐ MICROSCOPIC ☐

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

TERMINAL ILEUM	<input type="checkbox"/>	CAECUM	<input type="checkbox"/>	ASCENDING COLON	<input type="checkbox"/>
TRANSVERSE COLON	<input type="checkbox"/>	DESCENDING COLON	<input type="checkbox"/>	SIGMOID COLON	<input type="checkbox"/>
RECTUM	<input type="checkbox"/>				

8.6) Has the patient has ever been investigated for dysplasia? Yes ☐ No ☐

If no, please go to question 8.10.

8.7) 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 ☐

8.8) What was the endoscopic appearance of the dysplasia?

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

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

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

If no, please go to Q8.14

8.11) If yes to Q8.10, what was the date of their colectomy (if known)? (DD/MM/YYYY)

8.12) Please specify the type of colectomy (if known):

Sub-total colectomy with ileo-anal pouch ☐ Pan-proctocolectomy with ileostomy ☐

8.13) What was the indication for the colectomy?

Active disease ☐ Active disease and Neoplasia ☐ Low grade dysplasia ☐

High grade dysplasia ☐ Adenocarcinoma ☐ Other ☐

Unknown ☐

8.14) If patient has had colon cancer, please list the site

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

Yes ☐ No ☐

8.16) If yes, which treatments were they?

Anti-TNF ☐ Vedolizumab ☐

Other ☐ Please provide treatment name _____

Question 9

9.1) 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		

9.2) For patients having a cholecystectomy, please code the indication :

GB Cancer ☐ GB Polyp ☐ Cholelithiasis Related ☐ Unknown ☐

9.3) Has the patient had a bone density/DEXA scan? Yes ☐ No ☐

If no, proceed to Q.10

9.4) If yes, provide details of the most recent scan:

Date of the report (DD/MM/YYYY)

T score of hip..... T score of spine.....

Question 10

10.1) Please put the details of the **first** liver ultrasound findings the patient had:

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 ☐

Bile duct dilatation

Yes ☐No ☐

Lymph nodes at the porta

Yes ☐No ☐10.2) Please put the details of the **most recent** US the patient has had

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 11**

11.1) Does the patient suffer from any of the following co-morbid hepatic disorders?

	YES
CHRONIC HEPATITIS B	
CHRONIC HEPATITIS C	
NON-ALCOHOLIC FATTY LIVER DISEASE (NAFLD OR NASH)	
AUTOIMMUNE HEPATITIS (OR AUTOIMMUNE OVERLAP)	
PRIMARY BILIARY CHOLANGITIS	
HAEMOCHROMATOSIS	
OTHER CO-MORBID LIVER DISORDER (PLEASE SPECIFY)	

11.2) 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 12

12.1) Does the patient suffer from any of these additional diseases?

DISEASE	YES
TYPE 1 DIABETES	
PSORIASIS	
RHEUMATOID ARTHRITIS	
ANKYLOSING SPONDYLITIS	
SARCOIDOSIS	
MYAESTHENIA GRAVIS	
MULTIPLE SCLEROSIS	
COELIAC DISEASE	
SYSTEMIC LUPUS ERYTHEMATOSUS	
SJOGRENS SYNDROME	
HYPOTHYROIDISM	
HYPERTHYROIDISM	

Question 13

13.1) Has the patient ever taken any of the following medications?

MEDICATION	YES/NO	START DATE	END DATE	DOSE	INDICATION
URSODEOXYCOLIC ACID					
AZATHIOPRINE					
5-ASA COMPOUND					
PREDNISOLONE					
MMF					
CYCLOSPORINE					
TACROLIMUS					
OTHER MEDICATIONS					
OTHER BIOLOGICAL AGENTS FOR IBD					

13.2) What is the patient's most recent weight (please state units)?

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	<input type="checkbox"/>	CT report	<input type="checkbox"/>
MRCP Report	<input type="checkbox"/>	MRI report	<input type="checkbox"/>
Liver Biopsy Report	<input type="checkbox"/>	Hepatobiliary surgery report	<input type="checkbox"/>
Colonoscopy report	<input type="checkbox"/>	MRE report	<input type="checkbox"/>
Gastroscopy report	<input type="checkbox"/>	Cirrhotic morphology report	<input type="checkbox"/>

*If you have any **questions** or **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,
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Lv 6 Addenbrooke's Treatment Centre,
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