

A1. Site/Study ID#: _____ /IG _____

A2. Visit Date: _____ / _____ / _____
Month Day Year

A3. Staff Initials: _____

To DCC **SECTION B: INITIAL HISTORY OF PRESENT ILLNESS**

B1. On what date (month and year) did parents first notice symptoms of liver disease?

DATE **IHX2B01MM** V2(3)/ **IHX2B01YY** V2(4)/ **IHX2B01DT** 66. UNK **IHX2B01UK** V2(3)
Month Year

Check all symptoms that were present at time of first symptoms of liver disease:

- | | | | |
|---------------------------------------------------------------------------|---------------------------------|--------------------------------|---------------------------------|
| 1. Jaundice IHX2B011 V2(3) | 1. <input type="checkbox"/> Yes | 2. <input type="checkbox"/> No | 55. <input type="checkbox"/> DK |
| 2. Pruritus IHX2B012 V2(3) | 1. <input type="checkbox"/> Yes | 2. <input type="checkbox"/> No | 55. <input type="checkbox"/> DK |
| 3. Diarrhea IHX2B013 V2(3) | 1. <input type="checkbox"/> Yes | 2. <input type="checkbox"/> No | 55. <input type="checkbox"/> DK |
| 4. Failure to thrive IHX2B014 V2(3) | 1. <input type="checkbox"/> Yes | 2. <input type="checkbox"/> No | 55. <input type="checkbox"/> DK |
| 5. Bleeding or bruising IHX2B015 V2(3) | 1. <input type="checkbox"/> Yes | 2. <input type="checkbox"/> No | 55. <input type="checkbox"/> DK |
| 6. Ascites IHX2B016 V2(3) | 1. <input type="checkbox"/> Yes | 2. <input type="checkbox"/> No | 55. <input type="checkbox"/> DK |
| 7. GI bleeding IHX2B017 V2(3) | 1. <input type="checkbox"/> Yes | 2. <input type="checkbox"/> No | 55. <input type="checkbox"/> DK |
| 8. Bone fracture IHX2B018 V2(3) | 1. <input type="checkbox"/> Yes | 2. <input type="checkbox"/> No | 55. <input type="checkbox"/> DK |
| 9. Rickets IHX2B019 V2(3) | 1. <input type="checkbox"/> Yes | 2. <input type="checkbox"/> No | 55. <input type="checkbox"/> DK |
| 10. Vitamin E deficiency IHX2B110 V2(3) | 1. <input type="checkbox"/> Yes | 2. <input type="checkbox"/> No | 55. <input type="checkbox"/> DK |
| 11. Abnormal liver blood tests other than bilirubin IHX2B111 V2(3) | 1. <input type="checkbox"/> Yes | 2. <input type="checkbox"/> No | 55. <input type="checkbox"/> DK |

B2. On what date (month and year) was the diagnosis of this CLiC liver disease made?

DATE **IHX2B02MM** V2(3)/ **IHX2B02YY** V2(4)/ **IHX2B02DT** V2(3)/ 66. UNK **IHX2B02UK** V2(3)
Month Year

Check all symptoms that were present at time of diagnosis:

- | | | | |
|---------------------------------------------------------------------------|---------------------------------|--------------------------------|---------------------------------|
| 1. Jaundice IHX2B021 V2(3) | 1. <input type="checkbox"/> Yes | 2. <input type="checkbox"/> No | 55. <input type="checkbox"/> DK |
| 2. Pruritus IHX2B022 V2(3) | 1. <input type="checkbox"/> Yes | 2. <input type="checkbox"/> No | 55. <input type="checkbox"/> DK |
| 3. Diarrhea IHX2B023 V2(3) | 1. <input type="checkbox"/> Yes | 2. <input type="checkbox"/> No | 55. <input type="checkbox"/> DK |
| 4. Failure to thrive IHX2B024 V2(3) | 1. <input type="checkbox"/> Yes | 2. <input type="checkbox"/> No | 55. <input type="checkbox"/> DK |
| 5. Bleeding or bruising IHX2B025 V2(3) | 1. <input type="checkbox"/> Yes | 2. <input type="checkbox"/> No | 55. <input type="checkbox"/> DK |
| 6. Ascites IHX2B026 V2(3) | 1. <input type="checkbox"/> Yes | 2. <input type="checkbox"/> No | 55. <input type="checkbox"/> DK |
| 7. GI bleeding IHX2B027 V2(3) | 1. <input type="checkbox"/> Yes | 2. <input type="checkbox"/> No | 55. <input type="checkbox"/> DK |
| 8. Bone fracture IHX2B028 V2(3) | 1. <input type="checkbox"/> Yes | 2. <input type="checkbox"/> No | 55. <input type="checkbox"/> DK |
| 9. Rickets IHX2B029 V2(3) | 1. <input type="checkbox"/> Yes | 2. <input type="checkbox"/> No | 55. <input type="checkbox"/> DK |
| 10. Vitamin E deficiency IHX2B210 V2(3) | 1. <input type="checkbox"/> Yes | 2. <input type="checkbox"/> No | 55. <input type="checkbox"/> DK |
| 11. Abnormal liver blood tests other than bilirubin IHX2B211 V2(3) | 1. <input type="checkbox"/> Yes | 2. <input type="checkbox"/> No | 55. <input type="checkbox"/> DK |

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B3. Which of the following COMPLICATIONS have occurred in your child's lifetime: (indicate date of first appearance)

- | Complications | Date of First Appearance (MM/DD/YYYY) | | | |
|--------------------------------------------------------------------------------------------------------------------------|---------------------------------------|---------------------------------|--------------------------------|---------------------------------------------------------------------------------------------|
| 1. Cholangitis IHX2B031 V2(3)
IHX2B31MM V2(3)/ IHX2B31YY V2(4)/ IHX2B31DT | | 1. <input type="checkbox"/> Yes | 2. <input type="checkbox"/> No | 55. <input type="checkbox"/> DK
66. <input type="checkbox"/> UNK IHX2B31UN V2(3) |
| 2. Ascites IHX2B032 V2(3)
IHX2B32MM V2(3)/ IHX2B32YY V2(4)/ IHX2B32DT | | 1. <input type="checkbox"/> Yes | 2. <input type="checkbox"/> No | 55. <input type="checkbox"/> DK
66. <input type="checkbox"/> UNK IHX2B32UN V2(3) |
| 3. Bacterial Peritonitis IHX2B033 V2(3)
IHX2B33MM V2(3)/ IHX2B33YY V2(4)/ IHX2B33DT | | 1. <input type="checkbox"/> Yes | 2. <input type="checkbox"/> No | 55. <input type="checkbox"/> DK
66. <input type="checkbox"/> UNK IHX2B33UN V2(3) |
| 4. GI Bleed IHX2B034 V2(3)
IHX2B34MM V2(3)/ IHX2B34YY V2(4)/ IHX2B34DT | | 1. <input type="checkbox"/> Yes | 2. <input type="checkbox"/> No | 55. <input type="checkbox"/> DK
66. <input type="checkbox"/> UNK IHX2B34UN V2(3) |
| 5. Bone Fracture IHX2B035 V2(3)
IHX2B35MM V2(3)/ IHX2B35YY V2(4)/ IHX2B35DT | | 1. <input type="checkbox"/> Yes | 2. <input type="checkbox"/> No | 55. <input type="checkbox"/> DK
66. <input type="checkbox"/> UNK IHX2B35UN V2(3) |
| 6. Coagulopathy IHX2B036 V2(3)
IHX2B36MM V2(3)/ IHX2B36YY V2(4)/ IHX2B36DT | | 1. <input type="checkbox"/> Yes | 2. <input type="checkbox"/> No | 55. <input type="checkbox"/> DK
66. <input type="checkbox"/> UNK IHX2B36UN V2(3) |
| 7. Portopulmonary Hypertension IHX2B037 V2(3)
IHX2B37MM V2(3)/ IHX2B37YY V2(4)/ IHX2B37DT | | 1. <input type="checkbox"/> Yes | 2. <input type="checkbox"/> No | 55. <input type="checkbox"/> DK
66. <input type="checkbox"/> UNK IHX2B37UN V2(3) |
| 8. Hepatorenal Syndrome IHX2B038 V2(3)
IHX2B38MM V2(3)/ IHX2B38YY V2(4)/ IHX2B38DT | | 1. <input type="checkbox"/> Yes | 2. <input type="checkbox"/> No | 55. <input type="checkbox"/> DK
66. <input type="checkbox"/> UNK IHX2B38UN V2(3) |
| 9. Pruritus IHX2B039 V2(3)
IHX2B39MM V2(3)/ IHX2B39YY V2(4)/ IHX2B39DT | | 1. <input type="checkbox"/> Yes | 2. <input type="checkbox"/> No | 55. <input type="checkbox"/> DK
66. <input type="checkbox"/> UNK IHX2B39UN V2(3) |
| 10. Hearing problems IHX2B310 V2(3)
IHX2B310MM V2(3)/ IHX2B310YY V2(4)/ IHX2B310DT | | 1. <input type="checkbox"/> Yes | 2. <input type="checkbox"/> No | 55. <input type="checkbox"/> DK
66. <input type="checkbox"/> UNK IHX2B310UN V2(3) |
| 11. Rickets IHX2B311 V2(3)
IHX2B311MM V2(3)/ IHX2B311YY V2(4)/ IHX2B311DT | | 1. <input type="checkbox"/> Yes | 2. <input type="checkbox"/> No | 55. <input type="checkbox"/> DK
66. <input type="checkbox"/> UNK IHX2B311UN V2(3) |
| 12. Gallstones IHX2B312 V2(3) | | 1. <input type="checkbox"/> Yes | 2. <input type="checkbox"/> No | 55. <input type="checkbox"/> DK |

A1. Site/Study ID#: ____ /G _____

A2. Visit Date: ____ / ____ / ____
Month Day YearIHX2B312MM V2(3)/ IHX2B312YY V2(4)/ IHX2B312DT 66. UNK IHX2B312UN V2(3)13. Diarrhea IHX2B313 V2(3) 1. Yes 2. No 55. DKIHX2B313MM V2(3)/ IHX2B313YY V2(4)/ IHX2B313DT 66. UNK IHX2B313UN V2(3)14. Pancreatitis IHX2B314 V2(3) 1. Yes 2. No 55. DKIHX2B314MM V2(3)/ IHX2B314YY V2(4)/ IHX2B314DT 66. UNK IHX2B314UN V2(3)15. Transplant listing IHX2B315 V2(3) 1. Yes 2. No 55. DKIHX2B315MM V2(3)/ IHX2B315YY V2(4)/ IHX2B315DT 66. UNK IHX2B315UN V2(3)

16. Other (Specify: _ IHX2B316SP V2(300)_____)

IHX2B316MM V2(3)/ IHX2B316YY V2(4)/ IHX2B316DT 66. UNK IHX2B316UN V2(3)

17. Other (Specify: _ IHX2B317SP V2(300)_____)

IHX2B317MM V2(3)/ IHX2B317YY V2(4)/ IHX2B317DT 66. UNK IHX2B317UN V2(3)

18. Other (Specify: _ IHX2B318SP V2(300)_____)

IHX2B318MM V2(3)/ IHX2B318YY V2(4)/ IHX2B318DT 66. UNK IHX2B318UN V2(3)

19. Other (Specify: _ IHX2B319SP V2(300)_____)

IHX2B319MM V2(3) / IHX2B319YY V2(4)/ IHX2B319DT 66. UNK IHX2B319UN V2(3)

20. Other (Specify: _ IHX2B320SP V2(300)_____)

IHX2B320MM V2(3)/ IHX2B320YY V2(4)/ IHX2B320DT 66. UNK IHX2B320UN V2(3)

21. Other (Specify: _ IHX2B321SP V2(300)_____)

IHX2B321MM V2(3)/ IHX2B321YY V2(4)/ IHX2B321DT 66. UNK IHX2B321UN V2(3)B4. Were any abdominal surgeries performed? 1. Yes – Go to section CIHX2B04 V2(2) 2. Not - Done – Go to section D**SECTION C: INITIAL HISTORY OF ABDOMINAL SURGERY**C1. Was a drainage procedure performed on this subject during surgery? 1. Yes 2. No IHX2C01 V2(3)

A1. Site/Study ID#: _____ /G _____

A2. Visit Date: _____ / _____ / _____
Month Day Year

C2. Please identify the drainage procedure performed on this subject (choose all that apply):

Anatomic Region	Results	Type of Surgery
a. <input type="checkbox"/> Roux-en-kasai IHX2C02A V2(3)	Date: IHX2C02AMM V2(3)/ IHX2C02ADD V2(3)/ IHX2C02AYY V2(3)/ IHX2C02ADT (MM/DD/YYYY)	1. <input type="checkbox"/> Open 2. <input type="checkbox"/> Laparoscopic IHX2C02ATS V2(3)
b. <input type="checkbox"/> Gallbladder Kasai IHX2C02B V2(3)	Date: IHX2C02BMM V2(3)/ IHX2C02BDD V2(3)/ IHX2C02BYY V2(3)/ IHX2C02BDT (MM/DD/YYYY)	1. <input type="checkbox"/> Open 2. <input type="checkbox"/> Laparoscopic IHX2C02BTS V2(3)
c. <input type="checkbox"/> Choledochojejunostomy IHX2C02C V2(3)	Date: IHX2C02CMM V2(3)/ IHX2C02CDD V2(3)/ IHX2C02CYY V2(3)/ IHX2C02CDT (MM/DD/YYYY)	1. <input type="checkbox"/> Open 2. <input type="checkbox"/> Laparoscopic IHX2C02CTS V2(3)
d. <input type="checkbox"/> Partial biliary diversion IHX2C02D V2(3)	Date: IHX2C02DMM V2(3)/ IHX2C02DDD V2(3)/ IHX2C02DYY V2(3)/ IHX2C02DDT (MM/DD/YYYY)	1. <input type="checkbox"/> Open 2. <input type="checkbox"/> Laparoscopic IHX2C02DTS V2(3)
e. <input type="checkbox"/> Ileal exclusion IHX2C02E V2(3)	Date: IHX2C02EMM V2(3)/ IHX2C02EDD V2(3)/ IHX2C02EYY V2(3)/ IHX2C02EDT (MM/DD/YYYY) Length of ileum excluded: IHX2C2ECM V2(5)_ cm	1. <input type="checkbox"/> Open 2. <input type="checkbox"/> Laparoscopic IHX2C02ETS V2(3)
f. <input type="checkbox"/> Cholecystectomy IHX2C02F V2(3)	Date: IHX2C02FMM V2(3)/ IHX2C02FDD V2(3)/ IHX2C02FYY V2(3)/ IHX2C02FDT (MM/DD/YYYY) Gallstones present? 1. <input type="checkbox"/> Yes 2. <input type="checkbox"/> No IHX2C2FGP V2(3)	1. <input type="checkbox"/> Open 2. <input type="checkbox"/> Laparoscopic IHX2C02FTS V2(3)
g. <input type="checkbox"/> Other: (specify) IHX2GTGO V2(3) IHX2C02GOSP V2(300)	Date: IHX2C02GMM V2(3)/ IHX2C02GDD V2(3)/ IHX2C02GYY V2(3)/ IHX2C02GDT (MM/DD/YYYY) Specify: IHX2C02GSP V2(300)	1. <input type="checkbox"/> Open 2. <input type="checkbox"/> Laparoscopic IHX2C02GTS V2(3)

THE NEXT SECTION IS FOR ALAGILLE SYNDROME PATIENTS ONLY**SECTION D: ADDITIONAL ALAGILLE SYNDROME HISTORY**D1. Does this participant have Alagille Syndrome? **IHX2D01** V2(3) 1. Yes 2. No - END

D2. Which of the following features of Alagille Syndrome have occurred in participant's lifetime:

1. **Cardiac** 1. Yes 2. No - Go to 2 **IHX2D021** V2(3)
- a. Peripheral pulmonary stenosis **IHX2D021A** V2(3) 1. Yes 2. No 55. DK
- b. Pulmonary valve stenosis **IHX2D021B** V2(3) 1. Yes 2. No 55. DK
- c. Tetralogy of Fallot **IHX2D021C** V2(3) 1. Yes 2. No 55. DK
- d. Ventricular septal defect **IHX2D021D** V2(3) 1. Yes 2. No 55. DK
- e. Atrial septal defect **IHX2D021E** V2(3) 1. Yes 2. No 55. DK

A1. Site/Study ID#: ____ /G _____

A2. Visit Date: ____ / ____ / ____
Month Day Yearf. Other: **IHX2D021F** V2(3) **IHX2D21FSP** V2(300) 1. Yes 2. No 55. DK2. **Facies** 1. Yes 2. No – Go to 3 **IHX2D022** V2(3)a. Deep-set eyes **IHX2D022A** V2(3) 1. Yes 2. No 55. DKb. Broad forehead **IHX2D022B** V2(3) 1. Yes 2. No 55. DKc. Pointed chin **IHX2D022C** V2(3) 1. Yes 2. No 55. DK3. **Eyes** **IHX2D023** V(3) 1. Yes 2. No – Go to 4a. Posterior embryotoxon **IHX2D023A** V(3) 1. Yes 2. No 55. DKb. Axenfeld's anomaly **IHX2D023B** V(3) 1. Yes 2. No 55. DKc. Other: ____ **IHX2D023C** V(3) **IHX2D23CSP** V(300) ____ 1. Yes 2. No 55. DK4. **Skeletal** 1. Yes 2. No – Go to 5 **IHX2D024** V2(3)a. Butterfly vertebrae **IHX2D024A** V2(3) 1. Yes 2. No 55. DKb. Other: ____ **IHX2D024B** V(3) **IHX2D24BSP** V(300) ____ 1. Yes 2. No 55. DK5. **Renal** 1. Yes 2. No – Go to 6 **IHX2D025** V2(3)a. Dysplastic kidney **IHX2D025A** V2(3) 1. Yes 2. No 55. DKb. Single Kidney **IHX2D025B** V2(3) 1. Yes 2. No 55. DKc. Other structural anomaly: **IHX2D025C** V2(3) **IHX2D25CSP** V2(300) 1. Yes 2. No 55. DKd. Renal tubular acidosis **IHX2D025D** V2(3) 1. Yes 2. No 55. DK6. **Pancreas** 1. Yes 2. No – Go to 7 **IHX2D026** V2(3)a. Pancreatic insufficiency **IHX2D026A** V2(3) 1. Yes 2. No 55. DK7. **Vascular** 1. Yes 2. No – END **IHX2D027** V2(3)

a. Cerebrovascular accident (e.g., hemorrhage, thrombosis, aneurysm)

Specify: ____ **IHX2D027A** V2(3) **IHX2D27ASP** V2(300) ____ 1. Yes 2. No 55. DK

b. Stenosis or aneurysm of other blood vessels (e.g., renal artery stenosis, abdominal aneurysm, etc.)

Specify: **IHX2D027B** V2(3) **IHX2D27BSP** V2(300) 1. Yes 2. No 55. DK

6001

CLiC

Form 03.2 Group 2 - Initial History

A1. Site/Study ID#: ___ ___ /G ___ ___ ___

A2. Visit Date: ___ ___ / ___ ___ / ___ ___ ___
Month Day Year

IHX2CMMNT Comment V2(800)