SECTION A: GENERAL STUDY INFORMATION FOR OFFICE USE ONLY:

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

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

A3. Study Staff ID/Initials: _____ _____ _____

To DCC ☐

SECTION B: INFANT CHOLESTASIS

B1. Date of birth: _____ _____ / _____ _____ / _____ _____ _____ _____
Month Day Year

B2. Patient's sex: 1.☐ Male 2.☐ Female

B3. Is the patient Hispanic or Latino?

1.☐ Yes 2.☐ No 9.☐ Unknown

B4. Patient's racial background (check all that apply):

a. ☐ American Indian or Alaska Native
b. ☐ Asian
c. ☐ Black or African American
d. ☐ Native Hawaiian or Other Pacific Islander
e. ☐ White
f. ☐ Other (Specify: ___________________________________)
g. ☐ Unknown

SECTION A: GENERAL STUDY INFORMATION FOR OFFICE USE ONLY:

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

B5. Date of presentation to your center for cholestasis: _____ _____ / _____ _____ / _____ _____ _____ _____
Month Day Year

B6. Did the patient reportedly have pale, lightly pigmented, or acholic stools? 1.☐ Yes 2.☐ No 9.☐ NA

B7. Did the patient reportedly have hepatomegaly or a liver palpable more than 4 cm (3 finger-widths) below the costal margin? 1.☐ Yes 2.☐ No 9.☐ NA

B8. Did the patient reportedly have splenomegaly or a palpable spleen? 1.☐ Yes 2.☐ No 9.☐ NA

B9. Does the patient have any reported major congenital anomalies? 1.☐ Yes 2.☐ No 9.☐ NA

a. If "Yes," please select all that apply from the following list. For starred items, please provide specifics in B9b below.

ai. ☐ Polysplenia

aii. ☐ Asplenia

aiii. ☐ Situs abnormality

aiv. ☐ Intestinal malrotation

av. ☐ Preduodenal portal vein

avi. ☐ Interrupted inferior vena cava (IVC)

avii. ☐ Aberrant hepatic artery

aviii. ☐ Cardiac anomaly*

aix. ☐ Noncardiac vascular anomaly*

ax. ☐ Gastrointestinal anomaly*

axi. ☐ Urinary tract anomaly*

axii. ☐ Renal anomaly*

axiii. ☐ Pulmonary anomaly*

axiv. ☐ Malformation of an extremity

axv. ☐ Cleft lip

axvi. ☐ Cleft palate

axvii. ☐ Anomaly of the genitalia*

axviii. ☐ Other:_______________________________________

b. Please provide specifics on any starred items from B9a above:

____

____

____

____

____

____

____

SECTION A: GENERAL STUDY INFORMATION FOR OFFICE USE ONLY:

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

SECTION C: Diagnostic evaluations prior to hepatoportoenterostomy – specify all abnormalities if procedure was repeated

C1. Ultrasound:

a. Date of (last) Ultrasound: _____ _____ / _____ _____ / _____ _____ _____ _____ (Month / Day / Year) 9.☐ ND → **Skip to C2**

Anatomic Region	Results
b. Gallbladder:	**(Please check all that apply)** bi. ☐ Normal bii. ☐ Absent biii. ☐ Small/contracted biv. ☐ Irregular bv. ☐ Other:______________________ bvi. ☐ No information given
c. Extrahepatic Bile Duct:	**(Please check all that apply)** ci. ☐ Normal cii. ☐ Not visualized ciii. ☐ Cyst civ. ☐ Dilated cv. ☐ Other:______________________ cvi. ☐ No information given
d. Liver:	**(Please check all that apply)** di. ☐ Normal dii. ☐ Triangular cord diii. ☐ Intrahepatic cyst div. ☐ Other:______________________ dv. ☐ No information given
e. Other Findings:	**(Please check all that apply)** ei. ☐ Asplenia eii. ☐ Polysplenia eiii. ☐ Situs abnormality eiv. ☐ Preduodenal portal vein ev. ☐ Interrupted IVC evi. ☐ Other:______________________ evii. ☐ None

SECTION A: GENERAL STUDY INFORMATION FOR OFFICE USE ONLY:

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

C2. HIDA or Equivalent:

a. Date of (last) HIDA: _____ _____ / _____ _____ / _____ _____ _____ _____ (Month / Day / Year) 9.☐ ND → **Skip to C3**

b. Total time of study: _____ _____ hours

c. Did the patient receive phenobarbitol pretreatment? 1.☐ Yes 2.☐ No 9.☐ Unknown

d. Did the patient receive ursodeoxycholic acid pretreatment? 1.☐ Yes 2.☐ No 9.☐ Unknown

	Anatomic Region	Results
e.	Tracer uptake:	**(Please check all that apply)** ei. ☐ Normal eii. ☐ Reduced eiii. ☐ Enhanced eiv. ☐ Other:________ ev. ☐ No information given
f.	Tracer excretion:	**(Please check all that apply)** fi. ☐ Normal (visualized in bowel) fii. ☐ Gallbladder only fiii. ☐ Reduced or diminished fiv. ☐ Other:________ fv. ☐ None fvi. ☐ No information given

C3. MRCP:

a. Date of (last) MRCP: _____ _____ / _____ _____ / _____ _____ _____ _____ (Month / Day / Year) 9.☐ ND → **Skip to C4**

	Anatomic Region	Results
b.	Extrahepatic bile duct:	**(Please check all that apply)** bi. ☐ Normal bii. ☐ Not visualized biii. ☐ Cyst biv. ☐ Dilated bv. ☐ Other:________ bvi. ☐ No information given
c.	Liver:	**(Please check all that apply)** ci. ☐ Normal cii. ☐ Triangular cord ciii. ☐ Intrahepatic cyst civ. ☐ Other:________ cv. ☐ No information given

SECTION A: GENERAL STUDY INFORMATION FOR OFFICE USE ONLY:

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

C4. ERCP:

a. Date of (last) ERCP: _____ _____ / _____ _____ / _____ _____ _____ _____ (Month / Day / Year) 9.☐ ND → **Skip to C5**

	Results
b. Findings:	**(Please check all that apply)** bi. ☐ Normal bii. ☐ Normal pancreatogram only biii. ☐ Failure of cannulation biv. ☐ Other:________________ bv. ☐ No information given
c. Extrahepatic bile duct:	**(Please check all that apply)** ci. ☐ Normal cii. ☐ Not visualized ciii. ☐ Cyst civ. ☐ Dilated cv. ☐ Hypoplastic cvi. ☐ Irregular cvii. ☐ Other:________________ cviii. ☐ No information given

SECTION A: GENERAL STUDY INFORMATION FOR OFFICE USE ONLY:

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

C5. How many liver biopsies were performed pre-hepatoportoenterostomy on this infant? _____

C6. First Biopsy:

a. Date of biopsy: _____ _____ / _____ _____ / _____ _____ _____ _____ (Month / Day / Year) 9.☐ ND → **Skip to C9**

b. Type of biopsy: 1.☐ Wedge 2.☐ Needle

c. Findings reported (check all that apply):

- ci. Normal..☐
- cii. Cholestasis..☐
- ciii. Bile duct proliferation...............................☐
- civ. Cirrhosis..☐
- cv. Fibrosis..☐
- cvi. Giant cells..☐
- cvii. Bile duct plugs..☐
- cviii. Inflammation...☐
- cix. Ductal plate malformation ....☐
- cx. Other:___________________________☐
- cxi. No information given ....☐

C7. Second Biopsy:

a. Date of biopsy: _____ _____ / _____ _____ / _____ _____ _____ _____ (Month / Day / Year) 9.☐ ND → **Skip to C9**

b. Type of biopsy: 1.☐ Wedge 2.☐ Needle

c. Findings reported (check all that apply):

- ci. Normal..☐
- cii. Cholestasis..☐
- ciii. Bile duct proliferation...............................☐
- civ. Cirrhosis..☐
- cv. Fibrosis..☐
- cvi. Giant cells..☐
- cvii. Bile duct plugs..☐
- cviii. Inflammation...☐
- cix. Ductal plate malformation ....☐
- cx. Other:___________________________☐
- cxi. No information given ....☐

SECTION A: GENERAL STUDY INFORMATION FOR OFFICE USE ONLY:

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

C8. Third Biopsy:

a. Date of biopsy: _____ _____ / _____ _____ / _____ _____ _____ _____ (Month / Day / Year) 9.☐ ND → **Skip to C9**

b. Type of biopsy: 1.☐ Wedge 2.☐ Needle

c. Findings reported (check all that apply):

- ci. Normal...☐
- cii. Cholestasis...☐
- ciii. Bile duct proliferation...............................☐
- civ. Cirrhosis...☐
- cv. Fibrosis..☐
- cvi. Giant cells...☐
- cvii. Bile duct plugs..☐
- cviii. Inflammation..☐
- cix. Ductal plate malformation ....☐
- cx. Other:____________________________☐
- cxi. No information given ....☐

SECTION A: GENERAL STUDY INFORMATION FOR OFFICE USE ONLY:

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

C9. Hepatoportoenterostomy:

a. Date of hepatoportoenterostomy: _____ _____ / _____ _____ / _____ _____ _____ _____
Month Day Year

b. Were corticosteroids used in hospital after hepatoportoenterostomy? 1.☐ Yes 2.☐ No →**SKIP TO C9c**

bi. Number of days used: _____ _____ days

bii. Describe dosing regimen: ______________________________

c. Surgical/Pathologic findings (in report):

ci. Portal tract bile duct size: _____ _____ _____ microns 9.☐ NA

cii. Cirrhosis? 1.☐ Yes 2.☐ No 9.☐ NA

ciii. Giant cell transformation? 1.☐ Yes 2.☐ No 9.☐ NA

d. Date of hospital discharge (or death): _____ _____ / _____ _____ / _____ _____ _____ _____
Month Day Year

e. Medications at discharge (check all that apply):

ei. ☐ Ursodeoxycholic acid
eii. ☐ Corticosteroid
eiii. ☐ Vitamin A
eiv. ☐ Vitamin D
ev. ☐ Vitamin E
evi. ☐ TPGS vitamin E (liqui-E)
evii. ☐ Vitamin K
eviii. ☐ ADEK or equivalent
eix. ☐ MVI
ex. ☐ Antibiotic (Specify:______________________________)
exi. ☐ Other:______________________________
exii. ☐ Other:______________________________
exiii. ☐ Other:______________________________
exiv. ☐ Other:______________________________

SECTION A: GENERAL STUDY INFORMATION FOR OFFICE USE ONLY:

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

SECTION D: Growth, Laboratory Values and Medications – DISCONTINUE AT TIME OF OLT OR DEATH

GROWTH	Presen-tation (a)	HPE (b)	Dis-charge (c)	First PO Visit (d)	3 MO PO (e)	6 MO PO (f)	Age 12 MO (g)	Age 18 MO (h)	Age 24 MO, transplant or death (i)
D1. Date or NA									
D2. Height (cm)									
D3. Weight (kg)									
D4. Head circumference (cm)									
For TPN and nasogastric feeding, please check "Y" if it was used at any time from the last clinic visit to the current clinic visit.									
D5. TPN?	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N
D6. Ng fed?	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N

SECTION A: GENERAL STUDY INFORMATION FOR OFFICE USE ONLY:

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

LAB VALUES	Presen-tation (a)	HPE (b)	Dis-charge (c)	First PO Visit (d)	3 MO PO (e)	6 MO PO (f)	Age 12 MO (g)	Age 18 MO (h)	Age 24 MO, transplant or death (i)
Use data closest to the specified time point. Do not duplicate columns. HPE = labs as close to but prior to the date of HPE.									
D7. Date or NA									
D8. Total bilirubin (mg/dl)									
D9. Direct bilirubin * (mg/dl)									
D10. Conjugated bilirubin (mg/dl) *									
D11. ALT (U/L)									
D12. Alk phos (U/L)									
D13. GGTP (U/L)									
D14. Albumin (g/L)									
D15. Na (mEq/L)									
D16. Hematocrit (%)									
D17. WBC ($10^3/mm^3$)									
D18. Platelets ($10^3/mm^3$)									
D19. Prothrombin time (sec)									
D20. INR									
D21. Cholesterol (mg/dl)									
D22. Vitamin A (mcg/dl)									
D23. Vitamin E (mg/L)									
D24. Vitamin D (25OH in ng/ml)									
D25. Vitamin D (1.25-OH in pg/ml)									

***NOTE:** Answer either D9 OR D10, both are not required.

SECTION A: GENERAL STUDY INFORMATION FOR OFFICE USE ONLY:

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

MEDICATIONS	Presen-tation (a)	HPE (b)	Dis-charge (c)	First PO Visit (d)	3 MO PO (e)	6 MO PO (f)	Age 12 MO (g)	Age 18 MO (h)	Age 24 MO, transplant or death (i)
Please check "Yes" if a medication was used/prescribed at the time of the clinic visit. If the answer is "Yes" to any of the medications below, record the daily dose only, do not calculate dose per kg.									
D26. Date or NA									
D27. Vitamin K Daily Dose (mg):	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N
D28. Vitamin A Daily Dose (IU):	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N
D29. Vitamin D Daily Dose (IU):	☐ D3 ☐ D25OH ☐ D1,25OH ☐	☐ D3 ☐ D25OH ☐ D1,25OH ☐ None	☐ D3 ☐ D25OH ☐ D1,25OH ☐ None	☐ D3 ☐ D25OH ☐ D1,25OH ☐ None	☐ D3 ☐ D25OH ☐ D1,25OH ☐ None	☐ D3 ☐ D25OH ☐ D1,25OH ☐ None	☐ D3 ☐ D25OH ☐ D1,25OH ☐ None	☐ D3 ☐ D25OH ☐ D1,25OH ☐ None	☐ D3 ☐ D25OH ☐ D1,25OH ☐ None
D30. Vitamin E Daily Dose (IU):	☐ E ☐ TPGS E ☐ None	☐ E ☐ TPGS E ☐ None	☐ E ☐ TPGS E ☐ None	☐ E ☐ TPGS E ☐ None	☐ E ☐ TPGS E ☐ None	☐ E ☐ TPGS E ☐ None	☐ E ☐ TPGS E ☐ None	☐ E ☐ TPGS E ☐ None	☐ E ☐ TPGS E ☐ None
D31. ADEK Daily Dose (ml):	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N
D32. Ursodeoxycholic acid Daily Dose (mg):	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N
D33a. Antibiotics Specify Other:	☐ TMP-SMX ☐ Amoxicillin ☐ Neomycin ☐ Other ☐ None	☐ TMP-SMX ☐ Amoxicillin ☐ Neomycin ☐ Other ☐ None	☐ TMP-SMX ☐ Amoxicillin ☐ Neomycin ☐ Other ☐ None	☐ TMP-SMX ☐ Amoxicillin ☐ Neomycin ☐ Other ☐ None	☐ TMP-SMX ☐ Amoxicillin ☐ Neomycin ☐ Other ☐ None	☐ TMP-SMX ☐ Amoxicillin ☐ Neomycin ☐ Other ☐ None	☐ TMP-SMX ☐ Amoxicillin ☐ Neomycin ☐ Other ☐ None	☐ TMP-SMX ☐ Amoxicillin ☐ Neomycin ☐ Other ☐ None	☐ TMP-SMX ☐ Amoxicillin ☐ Neomycin ☐ Other ☐ None
Daily Dose (mg):									
D33b. Antibiotics Specify Other:	☐ TMP-SMX ☐ Amoxicillin ☐ Neomycin ☐ Other ☐ None	☐ TMP-SMX ☐ Amoxicillin ☐ Neomycin ☐ Other ☐ None	☐ TMP-SMX ☐ Amoxicillin ☐ Neomycin ☐ Other ☐ None	☐ TMP-SMX ☐ Amoxicillin ☐ Neomycin ☐ Other ☐ None	☐ TMP-SMX ☐ Amoxicillin ☐ Neomycin ☐ Other ☐ None	☐ TMP-SMX ☐ Amoxicillin ☐ Neomycin ☐ Other ☐ None	☐ TMP-SMX ☐ Amoxicillin ☐ Neomycin ☐ Other ☐ None	☐ TMP-SMX ☐ Amoxicillin ☐ Neomycin ☐ Other ☐ None	☐ TMP-SMX ☐ Amoxicillin ☐ Neomycin ☐ Other ☐ None
Daily Dose (mg):									
D34. Spironalactone Daily Dose (mg):	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N
D35. Furosemide Daily Dose (mg):	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N
Report corticosteriod use in the interval leading to the date of the current visit. Number of days is reported as the number of days that corticosteriod was administered during that interval period.									
D36. Corticosteriod Number of Days	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N	☐ Y ☐ N
Total Dose (mg):									

SECTION A: GENERAL STUDY INFORMATION FOR OFFICE USE ONLY:

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

SECTION E: Chronology of Sentinel Events/Complications – Use additional pages if necessary (Page _____ of _____)

	Event (a)	**Hospitalization** Y / N / Unk	**Date** (MM/DD/YYYY) (b)	**Intervention** (report all used) (c)
E1.	_____	☐ Y ☐ N ☐ U	___ ___/ ___ ___/ ___ ___ ___ ___	_____, _____, _____, _____, _____
E2.	_____	☐ Y ☐ N ☐ U	___ ___/ ___ ___/ ___ ___ ___ ___	_____, _____, _____, _____, _____
E3.	_____	☐ Y ☐ N ☐ U	___ ___/ ___ ___/ ___ ___ ___ ___	_____, _____, _____, _____, _____
E4.	_____	☐ Y ☐ N ☐ U	___ ___/ ___ ___/ ___ ___ ___ ___	_____, _____, _____, _____, _____
E5.	_____	☐ Y ☐ N ☐ U	___ ___/ ___ ___/ ___ ___ ___ ___	_____, _____, _____, _____, _____
E6.	_____	☐ Y ☐ N ☐ U	___ ___/ ___ ___/ ___ ___ ___ ___	_____, _____, _____, _____, _____
E7.	_____	☐ Y ☐ N ☐ U	___ ___/ ___ ___/ ___ ___ ___ ___	_____, _____, _____, _____, _____
E8.	_____	☐ Y ☐ N ☐ U	___ ___/ ___ ___/ ___ ___ ___ ___	_____, _____, _____, _____, _____
E9.	_____	☐ Y ☐ N ☐ U	___ ___/ ___ ___/ ___ ___ ___ ___	_____, _____, _____, _____, _____
E10.	_____	☐ Y ☐ N ☐ U	___ ___/ ___ ___/ ___ ___ ___ ___	_____, _____, _____, _____, _____

Event Codes

A. Ascites
C. Cholangitis
E. Esophageal or gastric varices without bleeding
F. Esophageal or gastric variceal hemorrhage
G. Other GI bleeding
H. Hepatopulmonary syndrome
P. Peritonitis
S. Splenomegaly
T. Other significant bleeding
W. Other:______________________________
X. Other:______________________________
Y. Other:______________________________
Z. Other:______________________________

Intervention Codes:

10. Antibiotics
11. Band ligation
12. β-blockade
13. Diuretics
14. Endoscopy
15. Gastric acid inhibitor
16. Liver biopsy
17. Paracentesis
18. Red blood cell transfusion
19. Re-operation
20. Sclerotherapy
21. Steroids
22. Surgical shunt
23. TIPSS
24. Vasoconstrictive agent
25. Other:______________________________
26. Other:______________________________
27. Other:______________________________
28. Other:______________________________
29. Other:______________________________
30. Other:______________________________
31. Other:______________________________

SECTION A: GENERAL STUDY INFORMATION FOR OFFICE USE ONLY:

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

E11. Date of transplant listing: _____ _____ / _____ _____ / _____ _____ _____ _____ (Month / Day / Year) 9.☐ ND

E12. Date of initiation of living donor evaluation: _____ _____ / _____ _____ / _____ _____ _____ _____ (Month / Day / Year) 9.☐ ND

E13. Did the patient have a liver transplant? 1.☐ Yes 2.☐ No → **SKIP TO E14**

a. Date of liver transplant: _____ _____ / _____ _____ / _____ _____ _____ _____ (Month / Day / Year)

b. What type of liver was received (check all that apply)?

bi. ☐ Cadaveric whole

bii. ☐ Reduced

biii. ☐ Split

biv. ☐ LRD

E14. Last date of follow-up: _____ _____ / _____ _____ / _____ _____ _____ _____ (Month / Day / Year)

a. Was the patient alive? 1.☐ Yes →**END** 2.☐ No

b. Date of death: _____ _____ / _____ _____ / _____ _____ _____ _____ (Month / Day / Year)

c. Cause of death: 1.☐ Accident 2.☐ Primary cardiac death 3.☐ Natural 9.☐ Unknown

Investigator/Coordinator Signature:________________________________ Date:___ ___/___ ___/___ ___ ___ ___ (Month / Day / Year)