HE

Pediatric Acute Liver Failure

HOSPITALIZATION EVALUATION

04/01/2008
Version 1.3

FORM KEYS

Patient ID __ __- __ __ __ __ - __ __ __

Date enrolled __ __ - __ __ - __ __
(mm-dd-yy)

COMPLETION LOG
Data Collector ID _____
Initials

Data Collection __ __ - __ __ - __ __

Date Entered __ __ - __ __ - __ __

Date Verified __ __ - __ __ - __ __
MM DD YY

Fax completed form to 412-383-7151

Hospitalization Evaluation

SECTION I: DEMOGRAPHICS

1. Gender: ☐ Male ☐ Female
2. Date of birth __ __-__ __-__ __ __ __ (mm-dd-yyyy)
3. Is the patient Hispanic, Latino, or Latina?
   - ☐ No
   - ☐ Yes → 3.1 Specify origin:
     - ☐ 1 Cuban
     - ☐ 2 Mexican
     - ☐ 3 Puerto Rican
     - ☐ 4 Other: _____
4. With what race does the patient identify? (check all that apply)
   - ☐ White or Caucasian
   - ☐ Black or African-American
   - ☐ Asian
   - ☐ American Indian or Alaska Native
   - ☐ Native Hawaiian or other Pacific Islander
   - ☐ Other _____
5. Years of education completed: _____ ☐ N/A ☐ Unknown
6. Number of siblings (full or half brothers/sisters): _____

SECTION II: ADMISSION HISTORY

1. Initial hospital admission __ __ - __ __ - __ __ (mm-dd-yy)
   - 1a. Hospital transfer ☐ Yes → Date of transfer: __ __ - __ __ - __ __ (mm-dd-yy)
   - ☐ No
2. Date and time enrolled __ __ - __ __ - __ __ __ __ - __ __ (24-hour time)
3. Date of onset of jaundice __ __ - __ __ - __ __ (mm-dd-yy) ☐ N/A, patient not jaundiced
4. Symptoms that prompted patient or parent to seek medical attention

	Yes	No	Unk		Yes	No	Unk
Nausea/vomit	☐	☐	☐	Lethargy	☐	☐	☐
Abdominal pain	☐	☐	☐	Malaise	☐	☐	☐
Rash	☐	☐	☐	Fever	☐	☐	☐
Seizure	☐	☐	☐	Altered consciousness	☐	☐	☐
Jaundice	☐	☐	☐	Other, specify _____	☐	☐	☐

5. Alcohol use? ☐ Yes ☐ No ☐ Unk
6. Ecstasy use within 7 days of onset of symptoms? ☐ Yes ☐ No ☐ Unk

Hospitalization Evaluation

SECTION III: SYSTEM REVIEW & FAMILY HISTORY

	Patient			Mother			Father & Extended Family (Paternal & Maternal)			Sibling		
1. Cardiomyopathy	Y	N	U	Y	N	U	Y	N	U	Y	N	U
2. Cardiac arrhythmia	Y	N	U	Y	N	U	Y	N	U	Y	N	U
3. Heart disease	Y	N	U	Y	N	U	Y	N	U	Y	N	U
4. Seizure disorder	Y	N	U	Y	N	U	Y	N	U	Y	N	U
5. Myopathy	Y	N	U	Y	N	U	Y	N	U	Y	N	U
6. NASH (fatty liver)	Y	N	U	Y	N	U	Y	N	U	Y	N	U
7. Malignancy	Y	N	U	Y	N	U	Y	N	U	Y	N	U
8. Autoimmune disease	Y	N	U	Y	N	U	Y	N	U	Y	N	U
9. Unexplained liver disease				Y	N	U	Y	N	U	Y	N	U
10. Consanguinity				Y	N	U	Y	N	U			
11. HELLP syndrome				Y	N	U						
12. Severe pre-eclampsia				Y	N	U						
13. AFLP (acute fatty liver pregnancy)				Y	N	U						
14. SIDS										Y	N	U

15. Were maternal serologies performed? ☐ Yes ☐ No ☐ Unk

	Pos	Neg	ND	Unk		Pos	Neg	ND	Unk
15.1 HepA IgM	☐	☐	☐	☐	15.6 HepC Ab	☐	☐	☐	☐
15.2 HBsAg	☐	☐	☐	☐	15.7 Toxo	☐	☐	☐	☐
15.3 HBsAb	☐	☐	☐	☐	15.8 Rubella	☐	☐	☐	☐
15.4 HBcAb	☐	☐	☐	☐	15.9 CMV	☐	☐	☐	☐
15.5 HBeAg	☐	☐	☐	☐	15.10 Herpes	☐	☐	☐	☐

SECTION IV: MEDICATIONS

1. Drugs taken within last 6 months, prior to initial hospital admission (check all that apply):

	Yes	No	Unk		Yes	No	Unk
Anesthetics	☐	☐	☐	Immunosuppressives	☐	☐	☐
Anti TB agents	☐	☐	☐	NSAIDS	☐	☐	☐
Antibiotics	☐	☐	☐	OTC	☐	☐	☐
Anticonvulsants	☐	☐	☐	Psychiatric	☐	☐	☐
Diabetes meds	☐	☐	☐	Vitamins/nutritional sup.	☐	☐	☐
Estrogens	☐	☐	☐	Other, specify	☐	☐	☐
Herbals	☐	☐	☐	_____			

2. Medications in last 1 month prior to initial hospital admission (include toxins, herbs, mushrooms, OTC meds, vitamins, anesthetics)

Medication/Toxin Name	Office Use	Date last taken	Total Daily Dose		Duration	
	__ __	__ __-__ __-__ __	__ __ __ __ __	☐ mg ☐ µg	__ __	☐ days ☐ months
	__ __	__ __-__ __-__ __	__ __ __ __ __	☐ mg ☐ µg	__ __	☐ days ☐ months
	__ __	__ __-__ __-__ __	__ __ __ __ __	☐ mg ☐ µg	__ __	☐ days ☐ months
	__ __	__ __-__ __-__ __	__ __ __ __ __	☐ mg ☐ µg	__ __	☐ days ☐ months
	__ __	__ __-__ __-__ __	__ __ __ __ __	☐ mg ☐ µg	__ __	☐ days ☐ months
	__ __	__ __-__ __-__ __	__ __ __ __ __	☐ mg ☐ µg	__ __	☐ days ☐ months

3. Acetaminophen use? ☐ Yes ☐ No

3.1 Amount taken:

☐ Single dose →

3.1.1 Total dose: __ __ __ __ __ __ mg
3.1.2 Date taken: __ __ - __ __ - __ __
3.1.3 Time taken: __ __ : __ __

☐ Chronic use →

3.1.4 Average daily dose __ __ __ __ __ __ mg
3.1.5 Number of days taken: _____
3.1.6 Was a dose of >100 mg/kg taken during illness? ☐ Yes ☐ No
3.1.7 Reason taken:
☐ Fever ☐ Pain ☐ Other ____________________

3.2 Acetaminophen toxicity:
☐ Suicide attempt
☐ Accidental overdose
☐ Unknown
☐ N/A

SECTION V: PHYSICAL EXAM (at time of enrollment in registry)

Height: __ __ __.__ cm ☐ actual ☐ estimate

1 in = 2.54 cm
1 lb = 0.45 kg

Pulse: __ __ __ beats/min

Blood pressure: __ __ __ /__ __ __ mmHg

Weight: __ __ __.__ kgs

Temperature: __ __.__ °C

Hepatic coma grade: ☐ 0 ☐ I ☐ II ☐ III ☐ IV ☐ not assessable

PRISM score: __ __ Date assessed: __ __ - __ __ - __ __ (mm-dd-yy) ☐ not assessed

Does the patient have the following:

	Yes	No		Yes	No
Peripheral edema	☐	☐	Hepatomegaly	☐	☐
Capillary refill > 2 secs	☐	☐	Hyper-reflexia	☐	☐
Splenomegaly	☐	☐	Pupillary dilatation	☐	☐
Ascites	☐	☐	Delayed development	☐	☐
Jaundice	☐	☐			

SECTION VI: LABORATORY TESTS AND SEROLOGIES

Blood *Differential*		Not Done
PMN	__ __ %	☐
Lymphocytes	__ __ %	☐
Eosinophils	__ __ %	☐
Monos	__ __ %	☐
Kidney/Electrolytes		
Uric acid	__ __ . __ mg/dL	☐
Urine ketones (degree of positivity) - + ++ +++ ++++		☐
Liver		
Factor VII	__ __ %	☐
Factor V	__ __ %	☐
Alk Phos	__ __ __ __ IU/L	☐
Total protein	__ __ . __ gm/dL	☐
Amylase	__ __ __ __ IU/L	☐
CK	__ __ __ __ __ IU/L	☐
Arterial/Toxins		
St. BC	__ __ . __ mEq/L	☐
Urine for heavy metals?	☐ Pos ☐ Neg	☐
Urine toxin screen?	☐ Pos ☐ Neg	☐
If positive, indicate _____		
Acetaminophen level	__ __ __ mg/L	☐
Miscellaneous	**Pos** **Neg**	
Anti-smooth muscle	☐ ☐ 1:__ __ __	☐
ANA	☐ ☐ 1:__ __ __	☐
pANCA	☐ ☐	☐
Anti-LKM	☐ ☐ 1:__ __ __	☐

Miscellaneous		Not Done
Alpha-1-AT P_i type	__ __	☐
Alpha-1-AT level	__ __ __ mg/dL	☐
Serum copper	__ __ __ µg /ml	☐
Urine copper	__ __ __ __ µg/24 hr	☐
Ceruloplasmin	__ __ mg/dL	☐
Alpha-fetoprotein	__ __ __ __ __ __ ng/ml	☐
Ferritin	__ __ __ __ __ __ ng/ml	☐
Serum iron	__ __ __ µg/dL	☐
T_4	__ __ __ µg/dL	☐
TSH	__ __ __ __ __ __ IU/ml	☐
Cortisol	__ __ . __ µg/dL	☐
SLA IgG antibody	__ __ . __ units	☐
IgG quantitative	__ __ __ __ mg/dl	☐

Serologies	**Pos**	**Neg**	Not Done
Anti-HAV IgM	☐	☐	☐
HBsAg	☐	☐	☐
Anti-HBc	☐	☐	☐
Anti-HBc IgM	☐	☐	☐
HBeAg	☐	☐	☐
Anti-HBs	☐	☐	☐
HBV-DNA	☐	☐	☐
Anti-HDV	☐	☐	☐
Anti-HCV	☐	☐	☐
HCV-RNA	☐	☐	☐
Anti-HEV	☐	☐	☐
Anti-HIV	☐	☐	☐
B-hCG	☐	☐	☐

SECTION VII: VIRAL STUDIES (if positive result, record Source code)

Infectious Disease	Culture		Antigen		Antibody (IgM)		PCR		Hybridization	
Adenovirus	+ - ND	_	+ - ND	_	+ - ND	_	+ - ND	_	+ - ND	_
Paramyxovirus	+ - ND	_	+ - ND	_	+ - ND	_	+ - ND	_	+ - ND	_
Herpes simplex	+ - ND	_	+ - ND	_	+ - ND	_	+ - ND	_	+ - ND	_
HHV-6	+ - ND	_	+ - ND	_	+ - ND	_	+ - ND	_	+ - ND	_
CMV	+ - ND	_	+ - ND	_	+ - ND	_	+ - ND	_	+ - ND	_
EBV	+ - ND	_	+ - ND	_	+ - ND	_	+ - ND	_	+ - ND	_
Parvovirus	+ - ND	_	+ - ND	_	+ - ND	_	+ - ND	_	+ - ND	_
ECHO virus	+ - ND	_	+ - ND	_	+ - ND	_	+ - ND	_	+ - ND	_
Other _____	+ - ND	_	+ - ND	_	+ - ND	_	+ - ND	_	+ - ND	_
Other _____	+ - ND	_	+ - ND	_	+ - ND	_	+ - ND	_	+ - ND	_

Source codes: 1=blood 2=tracheal aspirate 3=urine 4=liver tissue 5=stool 6=naso-pharyngeal

SECTION VIII: INFECTIONS (Check if sample was tested and record all positive cultures and date of first positive sample)

Site (check if sample tested)	Date 1st positive sample	S aureus	S epid	S pneum	E coli	Klebsiella	Fungus	Other specify	Outcome (Resolved or Continuing)
☐ Blood	__ __-__ __-__ __	☐	☐	☐	☐	☐	☐	_____	R C
☐ Tracheal aspirate	__ __-__ __-__ __	☐	☐	☐	☐	☐	☐	_____	R C
☐ Urine	__ __-__ __-__ __	☐	☐	☐	☐	☐	☐	_____	R C
☐ Catheters	__ __-__ __-__ __	☐	☐	☐	☐	☐	☐	_____	R C
☐ Wounds	__ __-__ __-__ __	☐	☐	☐	☐	☐	☐	_____	R C
☐ Ascites	__ __-__ __-__ __	☐	☐	☐	☐	☐	☐	_____	R C

Anti-microbials Used	Anti-microbial code (office use)	Prophylaxis	Bowel Decontamination	Infection Therapy
		☐	☐	☐
		☐	☐	☐
		☐	☐	☐
		☐	☐	☐
		☐	☐	☐
_____	__ __ ___ __	☐	☐	☐

SECTION IX: METABOLIC STUDIES

	Abnormal			
	Diagnostic	**Non-specific**	**Normal**	**Not Done**
Urine organic acids	☐	☐	☐	☐
Serum amino acids	☐	☐	☐	☐
Urine reducing substances	☐	☐	☐	☐
Skin fibroblasts	☐	☐	☐	☐
Bile acid profile	☐	☐	☐	☐
Galactosemia screen	☐	☐	☐	☐
Acylcarnitine profile	☐	☐	☐	☐
Urine succinylacetone	☐	☐	☐	☐
Mitochondrial studies	☐	☐	☐	☐
Glycogen storage disease	☐	☐	☐	☐
Other _____	☐	☐	☐	☐

SECTION X: SCANS, MONITORING, LIVER BIOPSY

Test	Date Performed	Findings
☐ CT or MR of brain	__ __-__ __-__ __	☐ Normal ☐ Hemorrhage ☐ Herniation ☐ Edema
☐ CT of abdomen	__ __-__ __-__ __	☐ Normal ☐ Ascites ☐ Inhomogenious
☐ Ultrasound of abdomen	__ __-__ __-__ __	☐ Normal ☐ Ascites ☐ Hypoechoic liver Doppler performed? ☐ Yes ☐ No If Yes, ☐ Patent with normal flow ☐ Patent with reverse flow ☐ Vessel occluded
☐ EEG	__ __-__ __-__ __	☐ Normal ☐ Seizure ☐ Slowing
☐ Liver biopsy ↓ Electron Microscopy ☐ Yes ☐ No Method: ☐ Needle ☐ Transjugular ☐ Wedge (surgery)	__ __-__ __-__ __	☐ Steatosis ☐ Giant cell hepatitis ☐ Necrosis → ☐ Panlobular ☐ Centrilobular ☐ Focal ☐ Fibrosis → ☐ Portal ☐ Bridging → Cirrhosis ☐ Yes ☐ No
☐ Skin biopsy	__ __-__ __-__ __	☐ Normal ☐ Diagnostic ☐ Not diagnostic
☐ Muscle biopsy	__ __-__ __-__ __	☐ Normal ☐ Diagnostic ☐ Not diagnostic
☐ Bone marrow biopsy	__ __-__ __-__ __	☐ Normal ☐ Diagnostic ☐ Not diagnostic

SECTION XI: LIST STATUS

1. Ever listed for transplant:

☐ Yes →

1.1 Date of initial listing: __ __-__ __-__ __ (mm-dd-yy)

1.2 At listing: UNOS status _____ MELD/PELD score _____

1.3 Currently listed:

☐ Yes →
Current UNOS status: _____
Current MELD/PELD score: _____

☐ No →
Date removed from list __ __ - __ __ - __ __
Primary reason removed from list (check one):
☐ Improved ☐ Irreversible brain damage
☐ Sepsis ☐ Medically unsuitable
☐ Other _____

☐ No →

1.4 Primary reason not listed (check one):
☐ Not sick enough, too well ☐ Irreversible brain damage
☐ Sepsis ☐ Active substance abuse
☐ Inadequate social support ☐ Active psychiatric disease
☐ Medically unsuitable ☐ Other _____

SECTION XII: OUTCOME (at time of hospital discharge, transplant, or death)

☐ 1. Hospital discharge →

1.1 Date of discharge: __ __ - __ __ - __ __

1.2 Location of patient:
☐ Home
☐ Not home, specify location _____

☐ 2. Transplant →

2.1 Date of transplant: __ __-__ __-__ __

2.2 Type of transplant (check one):
☐ Whole ☐ Auxiliary ☐ Reduced ☐ Split liver
☐ Living-related ☐ Hepatocyte ☐ Other _____

2.3 ABO compatible liver? ☐ Yes ☐ No

2.4 Resected liver weight: __ __ __ __ gm

2.5 Resected liver histology (check all that apply)
☐ Necrosis ☐ Fibrosis ☐ Inflammation
☐ Steatosis ☐ Storage

☐ 3. Died →

3.1 Date of death: __ __-__ __-__ __

3.2 Major underlying cause of death: _____ (see codebook)

3.3 Autopsy performed: ☐ Yes ☐ No

Was parent/guardian approached to obtain consent to collect registry samples at time of autopsy? ☐ Yes ☐ No

Was consent obtained? ☐ Yes ☐ No

Hospitalization Evaluation

SECTION XIII: DIAGNOSIS

At Enrollment (check one)

☐ Acetaminophen ☐ Budd-Chiari ☐ Hemophagocytic syndrome
☐ Shock/ischemia ☐ Neonatal iron storage disease ☐ Veno-occlusive disease
☐ Metabolic liver disease →
- ☐ Respiratory chain deficit ☐ Tyrosinemia
- ☐ Wilson's disease ☐ Fatty acid oxidation _____
- ☐ Alpha-1-antitrypsin ☐ Mitochondrial _____
- ☐ Other _____

☐ Hepatitis →
- ☐ Viral: ☐ A ☐ B (±delta) ☐ C ☐ E ☐ EBV ☐ CMV ☐ Herpes simplex
  ☐ Other _____
- ☐ Autoimmune
- ☐ Drug-induced, agent _____

☐ Other _____
☐ Indeterminate

Final (check all that apply – rank if more than one)

☐ Acetaminophen ☐ Budd-Chiari ☐ Hemophagocytic syndrome
☐ Shock/ischemia ☐ Neonatal iron storage disease ☐ Veno-occlusive disease
☐ Metabolic liver disease →
- ☐ Respiratory chain deficit ☐ Tyrosinemia
- ☐ Wilson's disease ☐ Fatty acid oxidation _____
- ☐ Alpha-1-antitrypsin ☐ Mitochondrial _____
- ☐ Other _____

☐ Hepatitis →
- ☐ Viral: ☐ A ☐ B (±delta) ☐ C ☐ E ☐ EBV ☐ CMV ☐ Herpes simplex
  ☐ Other _____
- ☐ Autoimmune
- ☐ Drug-induced, agent _____

☐ Other _____
☐ Indeterminate

SECTION XIV: COMMENTS: ☐ Yes ☐ No

IF YES

_____
_____
_____
_____
_____
_____
_____