GENERAL INSTRUCTIONS

The Screening Evaluation (SE) form is completed for all patients who are younger than 18 years of age at the time of screening and show biochemical evidence of acute liver failure. Biochemical evidence of acute liver failure is defined as severe liver dysfunction, marked by elevated liver transaminases, occurring within 8 weeks of onset of illness, with no known underlying chronic liver disease.

The Screening Evaluation form must be completed for every patient clinically eligible for participation in the PALF Cohort study, regardless of whether or not they are enrolled in the study.

For example:

- A patient who meets all eligibility criteria but refuses consent must have a completed SE form.
- A patient who meets all eligibility criteria but patient consent is not obtained because you were not made aware of the patient, must have a completed SE form.

The Screening Evaluation form captures a minimal amount of demographic information along with the inclusion and exclusion criteria for determining eligibility, regardless of whether the patient is enrolled in the PALF Cohort study.

All patients screened for participation in the PALF Cohort study will be assigned a Screen ID. Only patients enrolled in the Cohort study will be assigned a Patient ID. Both IDs will be assigned by the data system.

SPECIFIC INSTRUCTIONS

Screen ID: A Screen ID is assigned by the PALF Data Entry System upon entry of the Screening Evaluation form. The Screen ID assigned by the system should be recorded on the data form, if a paper form is completed.

Section I: Screening Demographic

Date Screened: Record the month and two digit year that the patient is screened for participation in the PALF Cohort study. The screening date is the date that a clinically eligible patient is identified as potentially eligible for participation in the study.

Age: Record the age of the patient on the initial screening date and then check and check “Days”, “Months”, or “Years”. Use the following guidelines to record age:

- If less than 1 month old, record the age of the patient in days.
- If less than 5 years of age, record the age of the patient in months.
- For ages 5 and older, record the age in years,

Record -3 if the age of the patient at screening is unknown.

Sex: Check “Male” or “Female” to indicate the patient gender.

Hispanic: Check “Yes” or “No” to indicate whether or not the patient identifies as Hispanic, Latino, or Latina. Hispanic is defined as a person of Cuban, Mexican, Puerto Rican, South or Central American, or other Spanish culture or origin, regardless of race. The term “Spanish origin” can also be used in addition to “Hispanic or Latino”.

Hispanic, specify: If the patient identifies as Hispanic, mark the appropriate box that specifies origin.

If origin is not Cuban, Mexican, or Puerto Rican, check the “other” box and specify the place of origin given by the patient or parent/guardian.

If the patient has multiple Hispanic origins, check the "other" box and specify all of the origins in the text box.

If the patient identifies as Hispanic but the specific origin is unknown, check "unknown".

Race: Check the appropriate box to indicate the race of the patient. If the patient identifies with more than one race, check all that apply.

White or Caucasian: A person having origins in any of the original peoples of Europe, the Middle East, or North Africa.

Black or African-American: A person having origins in any of the black racial groups of Africa. Terms such as "Haitian" or "Negro" can be used in addition to "Black or African-American".

Asian: A person having origins in any of the original peoples of the Far East, Southeast Asia, or the Indian subcontinent including, for example, Cambodia, China, India, Japan, Korea, Malaysia, Pakistan, the Philippine Islands, Thailand, and Vietnam.

American Indian or Alaska Native: A person having origins in any of the original peoples of North, Central, or South America, and who maintains tribal affiliations or community attachment.

Native Hawaiian or Pacific Islander: A person having origins in any of the original peoples of Hawaii, Guam, Samoa, or other Pacific Islands.

Other: If the patient's racial background is not listed as one of the above options, check "Other" and specify the patient's race in the space provided.

Unknown: If the patient's racial background is not listed in the medical chart, the patient or parent/guardian did not identify the patient's race, and this information cannot be obtained via patient contact or a telephone call, check the "Unknown" box.

NOTE: Race must be provided regardless of ethnicity (Hispanic or not).

Section II: Inclusion Criteria

Inclusion Criteria: For each criterion, check "Yes" or "No" to indicate whether or not the patient meets the criterion. The response to all inclusion criteria must be YES for a patient to be eligible for participation in the study.
If a criterion is unknown or not assessed leave the response blank.

Consent: The patient or parent/guardian must be asked to provide written informed consent to participate in the study, using the most recent version of the IRB/REB approved consent document.

If the patient or parent/guardian does not consent to the study, indicate the reason.

Yes consent: Date approached: Record the date (month/day/year) that the patient or parent/guardian was initially approached for participation in the PALF cohort (i.e., when the study was first explained to the patient or parent/guardian or consent documents were offered to the patient or parent/guardian).

Date consent: Record the date (month/day/year) that the patient or parent/guardian provided consent for the PALF Cohort study (i.e., signed the consent documents).

INR/Encephalopathy: To meet this criterion, the patient must have an INR
a) greater than or equal to 1.5 and less than 2.0 with encephalopathy, or
b) greater than or equal to 2.0 with or without encephalopathy,
and uncorrected by Vitamin K.

If encephalopathy is not assessable because the patient is sedated, intubated, or on a ventilator, consider the encephalopathy criteria to be met.

Use the most recent INR and encephalopathy results performed prior to plasma therapy, but no more than 72 hours prior to enrollment into the cohort.

INR: Record the INR result and date of sample that was used to determine eligibility. If any part of the date is unknown, enter -3 for the unknown part of the date and enter the other parts of the date that are known. If the entire date is unknown, check "Unknown".

Encephalopathy: Record the encephalopathy score according to the age appropriate scale below, and the date of the assessment used to determine eligibility. If any part of the date is unknown, enter -3 for the unknown part of the date and enter the other parts of the date that are known. If the entire date is unknown, check "Unknown".

If the encephalopathy recorded in the hospital chart indicates a range rather than a single value; for example, a grade of 2-3, record the lower value.

If encephalopathy cannot be assessed (e.g., patient is sedated), check "Not assessable".

If encephalopathy could be assessed but is not assessed (e.g., doctor did not choose to assess), check "Not done".

For patients from **3 to 10 years of age** use the Standard Clinical Scales:

Stage	Clinical	Asterixis/Reflexes	Neurological signs	EEG changes
0	None	None/normal	Psych testing only	Normal
I	Confused, mood changes, altered sleep habits, loss of spatial orientation, forgetful	None/normal	Tremor, apraxia, impaired handwriting	Normal or diffuse slowing to theta rhythm, triphasic waves
II	Drowsy, inappropriate behavior, decreased inhibitions	None/ hyperreflexic	Dysarthria, ataxia	Abnormal generalized slowing, triphasic waves
III	Stuporous, obeys simple commands	None/hyperreflexia, up-going toes (+ Babinski)	Rigidity	Abnormal generalized slowing, triphasic waves
IV	Comatose, arouses with painful stimuli (IVa), or no response (IVb)	Absent	Decerebrate or decorticate	Abnormal, very slow delta activity

For patients **less than 3 years of age** use the Peter Whitington scale:

Stage	Clinical	Asterixis/Reflexes	Neurological signs
Early (I and II)	Inconsolable crying, sleep reversal, inattention to task	Unreliable/ normal or hyperreflexic	Untestable
Mid (III)	Somnolence, stupor, combativeness	Unreliable/hyperreflexic	Most likely untestable
Late (IV)	Comatose, arouses with painful stimuli (IVa) or no response (IVb)	Absent	Decerebrate or decorticate

Section III: Exclusion Criteria

Exclusion Criteria:

Check "Yes" or "No" to indicate if the patient meets any of the following exclusion criteria. The response to all exclusion criteria must be NO for a patient to be eligible for participation in this study. If a criterion is unknown or not assessed leave the response blank.

Known chronic liver disease: Any biochemical, diagnostic, or clinical evidence (in the opinion of the investigator) of any type of chronic underlying liver disease prior to enrollment.

Coagulopathy corrected with Vitamin K: If Vitamin K was given and corrected the coagulopathy, check "Yes." If the patient has not received Vitamin K or if coagulopathy persists despite treatment with Vitamin K (or another intervention intended to correct coagulopathy), check "No."

Multi-organ system failure: The patient is not eligible if they experience multi-organ system failure (MOSF) following heart surgery or ECMO, per the following definitions:

MOSF is the simultaneous presence of physiologic dysfunction or failure of two or more bodily organs or systems (cardiovascular, pulmonary, renal, neurological, hematological, hepatic or gastrointestinal).

Extracorporeal membrane oxygenation (ECMO) is a machine similar to a heart-lung machine; it pumps blood from the patient through a membrane oxygenator that imitates the gas exchange process of the lungs, i.e. it removes carbon dioxide and adds oxygen.

Prior transplant: A history of any solid organ (e.g., liver, kidney, pancreas, heart, lung, intestine, etc.) or bone marrow transplantation.

Acute trauma: Any sudden or rapid injury or trauma that, in the opinion of the investigator, would preclude participation in the study. Examples include: motor vehicle accident, fall, crushing injuries, sudden blows.

Previously enrolled: The patient is not eligible if they are already enrolled in the PALF Cohort study (funded from 2010-2015). If the patient was enrolled in the PALF registry (funded from 2005-2010) they are eligible to be enrolled in the PALF Cohort study.

Other Illness: Any other illness, condition, medical or other reason that would make the patient unsuitable for participation in the study, in the opinion of the investigator.

Enrollment:

All cohort inclusion criteria must be met and none of the exclusion criteria met for the patient to be eligible for enrollment into the PALF Cohort study.

Check “Yes” or “No” to indicate whether or not the patient is enrolled in the PALF cohort.

Date eligibility determined: Regardless of whether or not the patient is enrolled in the Cohort study, record the date (month/day/year) that eligibility is determined.

Not enrolled: Record the code associated with the initial primary diagnosis at the time of screening. The diagnosis is based on all information available at the time. The initial diagnosis should not reflect information obtained after eligibility is determined. If the diagnosis requires more information, specify in the space provided.

Enrolled: A Patient ID is assigned to eligible patients by the PALF Data Collection and Entry System upon entry of the Screening Evaluation form. The Patient ID assigned by the system should be recorded on the data form, if a paper form is completed.

The Patient ID is to be used as the participant identifier for all PALF forms.